EMBRYOLOGY OF HEART - 1

Dr. Anurag Bahekar

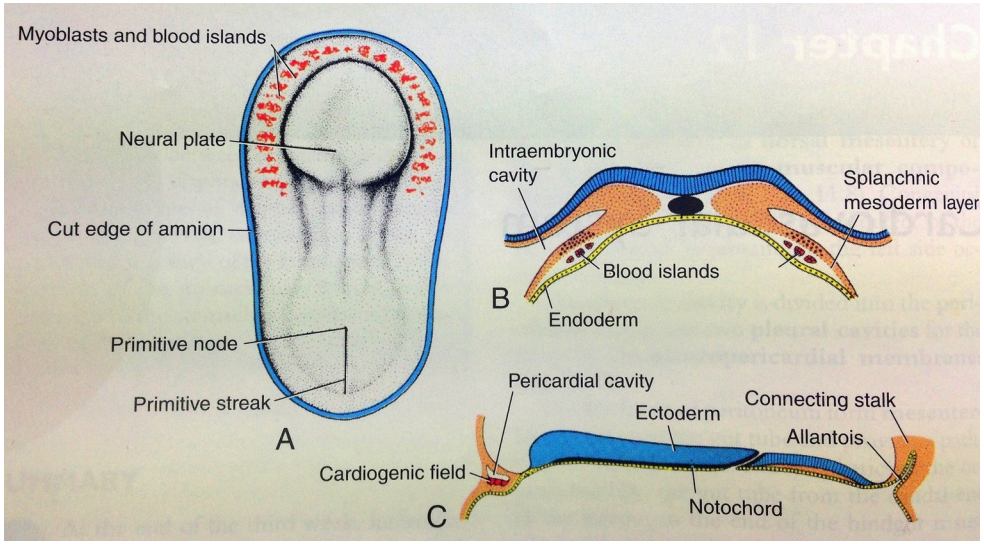
Headings covered

- Establishment of cardiogenic field
- Formation of heart tube
- Formation of cardiac loop

- Development of sinus venosus
- Atrial septa formation
- Differentiation of atria
- AV canal septal formation
- AV valves
- Conus & Truncus septa formation
- Ventricular septa formation
- Semilunar valves
- Conduction system

Establishment of cardiogenic field

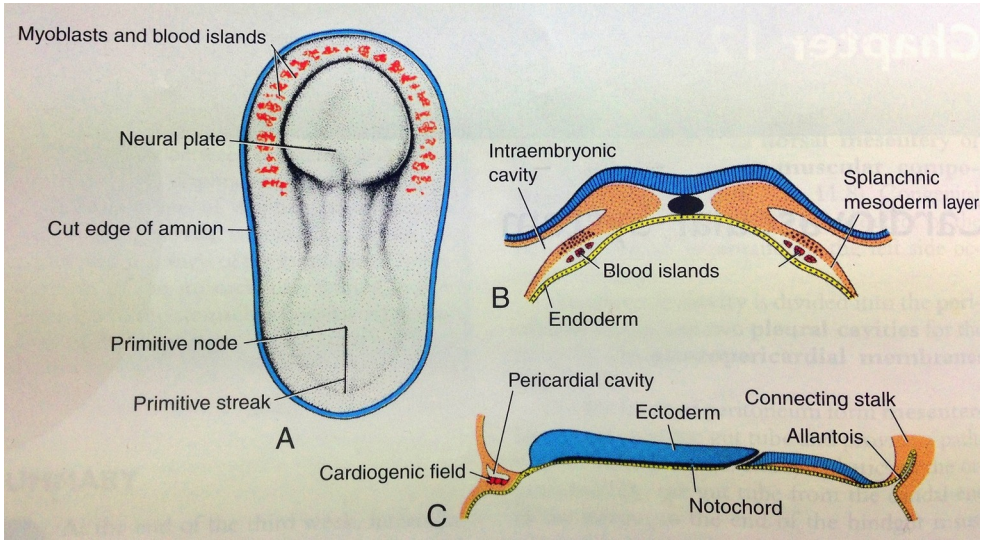
- Vascular system appears in middle of 3rd week
- Cardiac proginitor cells lie in the epiblast lateral to the primitive streak
- Migrate & reside in the splanchnic layer of the lateral plate mesoderm
- Induced by pharyngeal endoderm to form cardiac myoblasts



AT 18 DAYS

Establishment of cardiogenic field

- Blood islands appear in mesoderm
- Islands unite and form horse-shoe shaped endothelial-lined tube surrounded by myoblasts.
- This region is called **CARDIOGENIC FIELD**

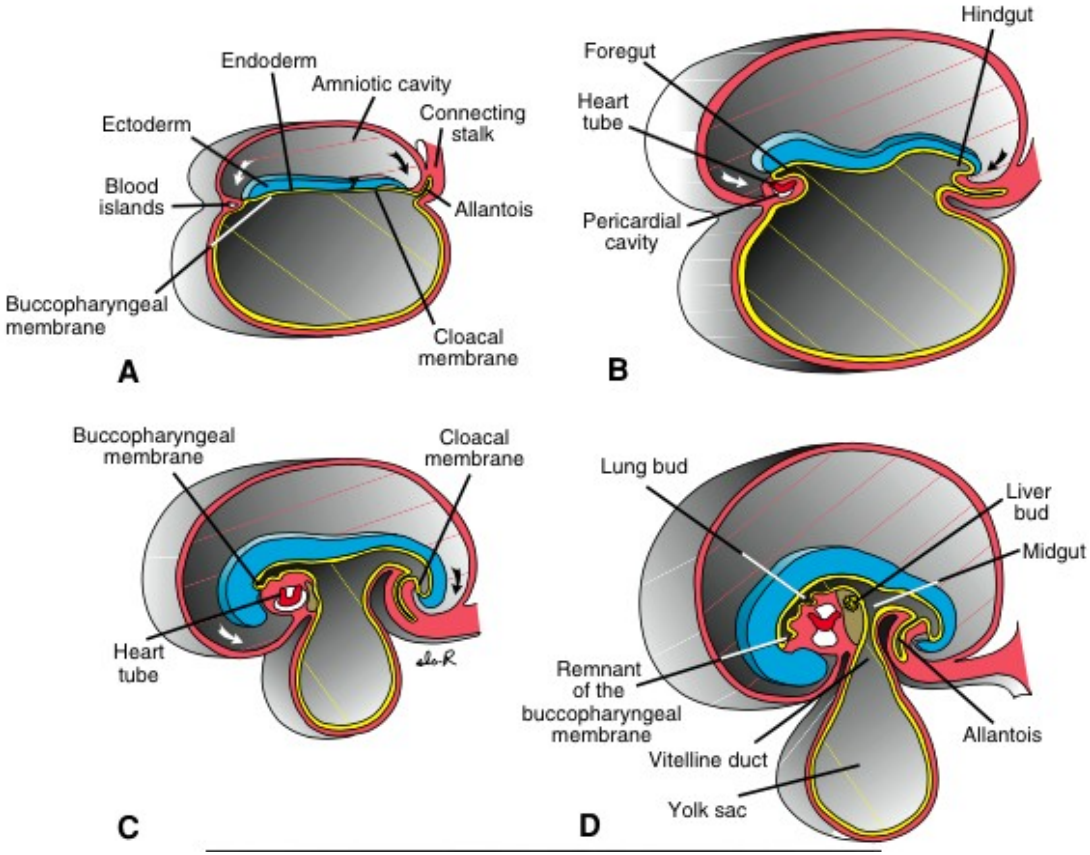


AT 18 DAYS

Andersons Paediatric Cardiology, Langman's Medical Embryology 10th Ed

Formation & position of heart tube

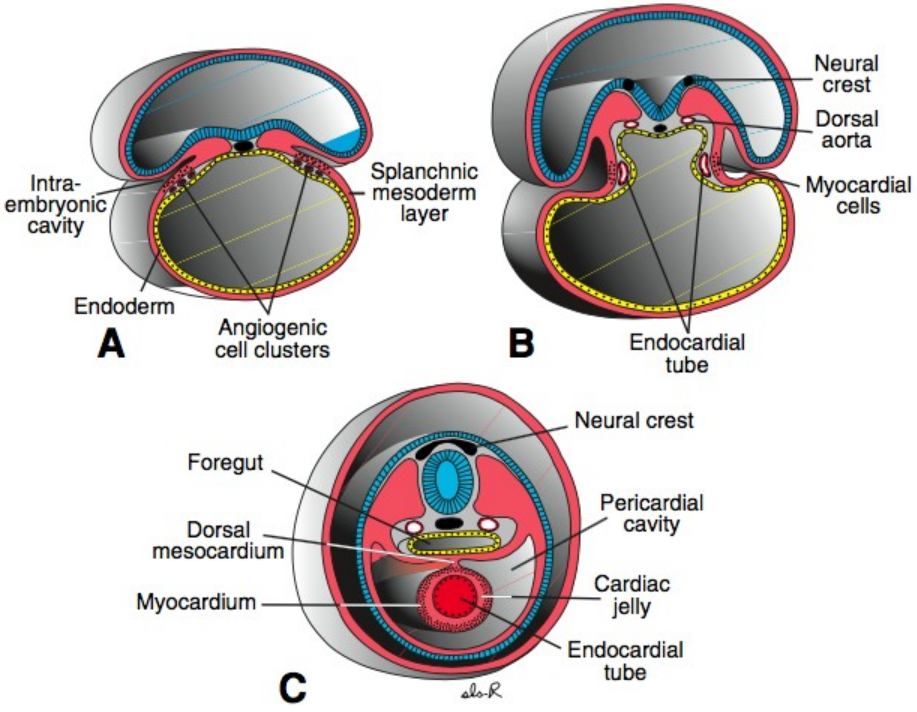
- Initially, cardiogenic area is anterior to the Buccopharyngeal membrane & neural plate
- CNS grows cephalad very rapidly
- Hence BP membrane is



Andersons Paediatric Cardiology, Langman's Medical Embryology 10th Ed

Formation & position of heart tube

- Part of wall of coelomic cavity becomes the myocardial muscle
- The dorsal mesocardium ensures the connection between extracoelomic mesenchyme and the heart tube
- Space between myocardial mantle & heart tube is filled with cardiac jelly



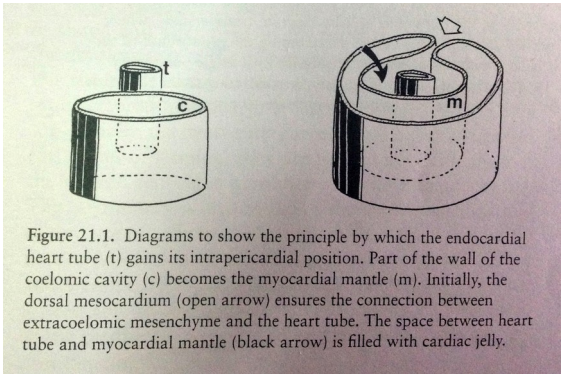
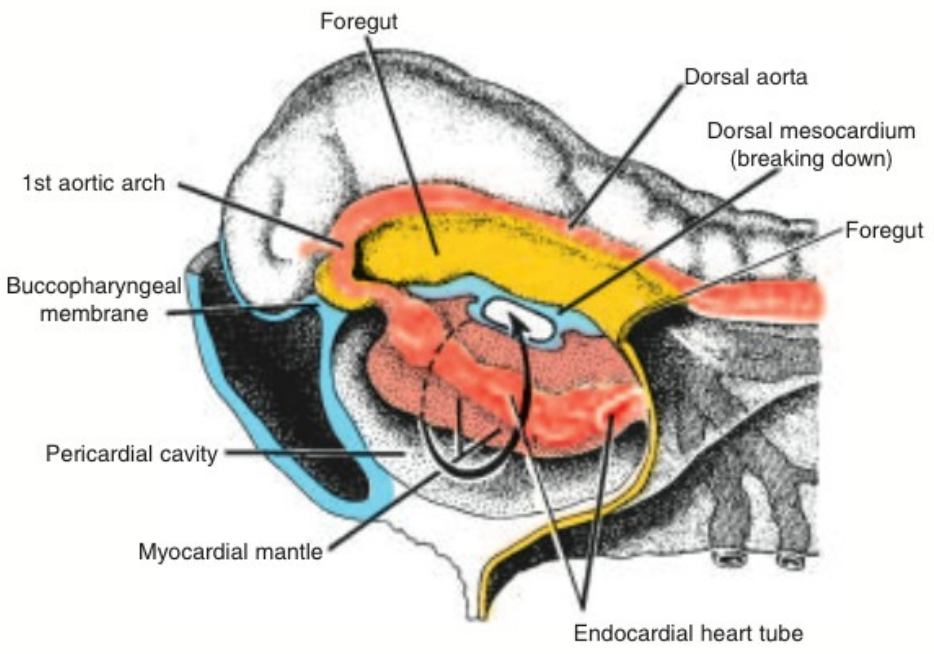


Figure 21.1. Diagrams to show the principle by which the endocardial heart tube (t) gains its intrapericardial position. Part of the wall of the coelomic cavity (c) becomes the myocardial mantle (m). Initially, the dorsal mesocardium (open arrow) ensures the connection between extracoelomic mesenchyme and the heart tube. The space between heart tube and myocardial mantle (black arrow) is filled with cardiac jelly.

Formation & position of heart tube

- Tube attached to dorsal side of pericardial cavity by a fold of DORSAL MESOCARDIUM

- Dorsal mesocardium – transverse pericardial sinus



Andersons Paediatric Cardiology, Langman's Medical Embryology 10th Ed

Development of heart

- **Myocardium** thickens and secretes ECM rich in hyaluronic acid

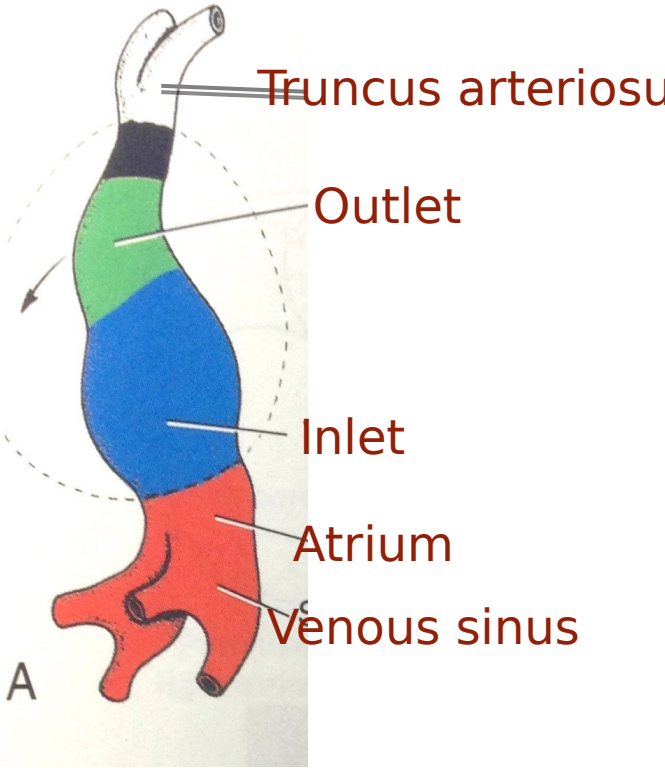
- **Epicardium** from mesothelial cells
  - on surface of septum transversum
  - in outflow tract region

- **Coronary artery** endothelium & smooth muscle from
  - Epicardial layer

Andersons Paediatric Cardiology,
Langman's Medical Embryology
10th Ed

Primitive cardiac segments

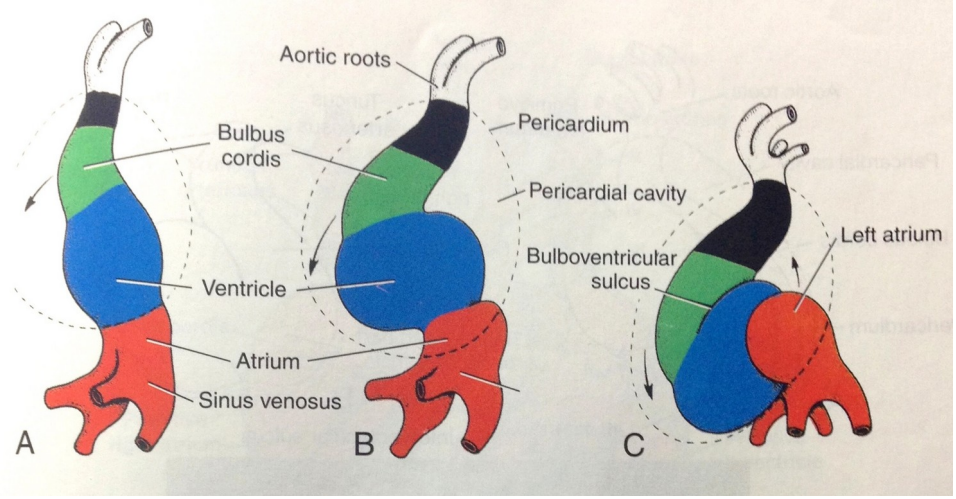
- On basis of constrictions in myocardial mantle
- Myocardium at junctions described as sphincters (Benninghoff)
- Within the heart these junctional zones are demarcated by "endocardial cushions" derived from cardiac jelly

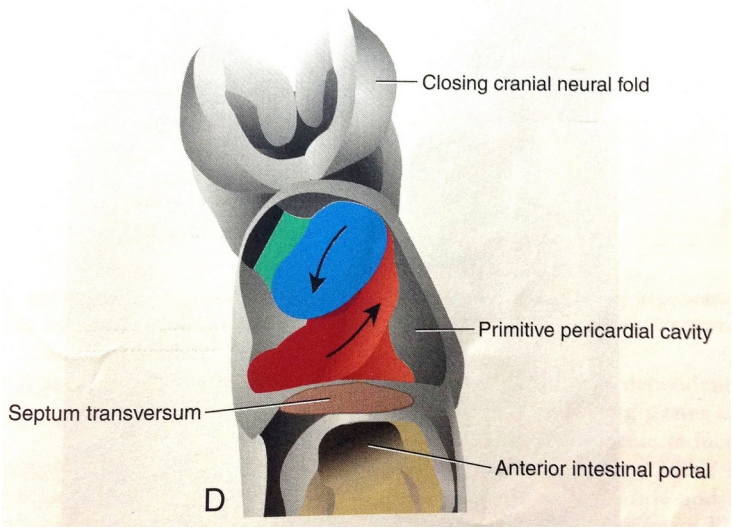


Andersons Paediatric Cardiology, Langman's Medical Embryology 10th Ed

Formation of cardiac loop

- Elongates and bends on **day 23**
- Cephalic portion :- Ventrally, caudally & to right
- Caudal portion :- Dorsocranially and to left
- **Due to cell shape changes**
- Organisation of these





Theories of cardiac looping

- Rapid development of tube within slowly developing pericardial cavity (Patten 1968)
- Haemodynamics as a morphogenic factor
- Changes in shape & alignment of myocardial cells (Manasek et al 1972)
- Multifactorial (Stallsberg 1970, Manasek 1981)

Andersons Paediatric Cardiology,
Langman's Medical Embryology
10th Ed

Abnormalities of cardiac looping

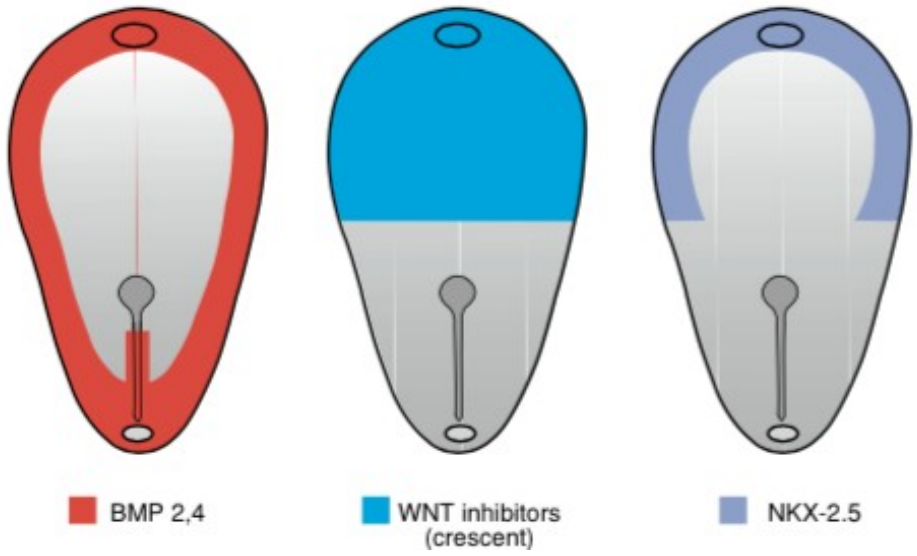
- Dextrocardia- heart loops to the left instead of right.
- Heterotaxy- sidedness is random

- Genes regulating sidedness are expressed during gastrulation
- Genes **nodal & lefty2** induce expression of transcription factor PITX2

- **PITX2** has role in deposition & function of ECM during looping.
- **NKX2** upregulates HAND1 & HAND2 that participate in looping of heart.

Molecular regulation

- Signals of **NKX2.5** (master gene for heart development) from anterior cranial endoderm induces a heart forming region in splanchnic mesoderm
- Signals require secretion of **BMP 2 & 4** by endoderm and lateral plate mesoderm
- WNT proteins inhibit heart development



Andersons Paediatric Cardiology, Langman's Medical Embryology 10th Ed

Clinical correlates

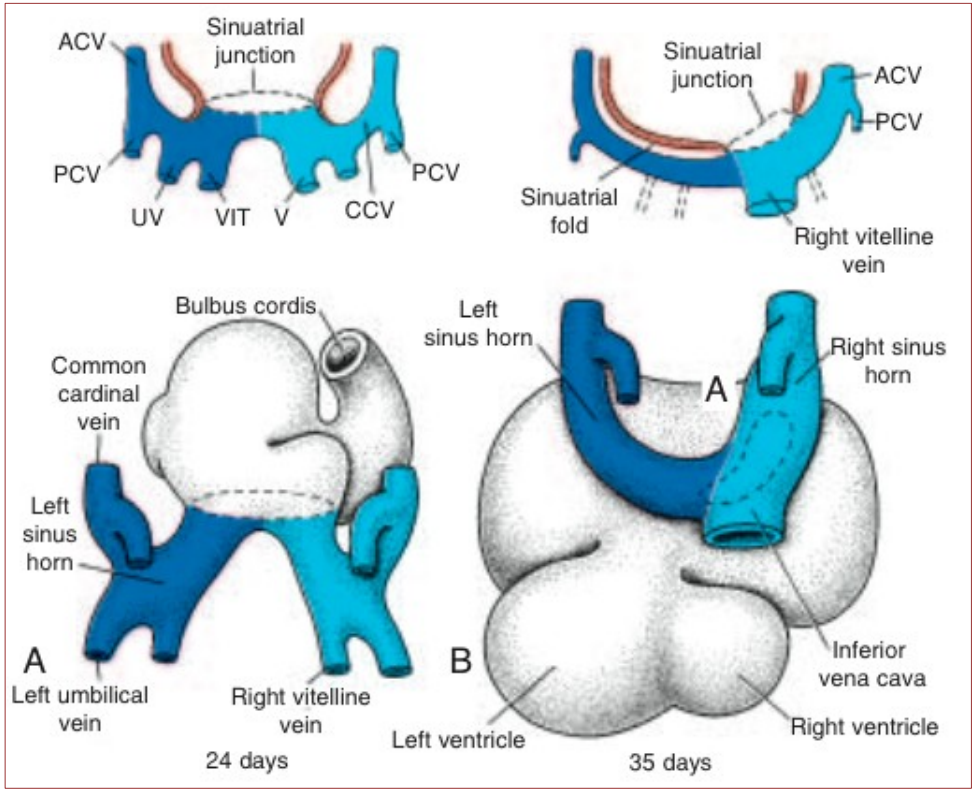
- **NKX2.5 mutations** on chromosome **5q35**- ASD, TOF, AV conduction delays in an AD fashion

- **TBX5 mutations** results in Holt Oram syndrome, Defects in muscular portions of IVS

Andersons Paediatric Cardiology,
Langman's Medical Embryology
10th Ed

Development of sinus venosus

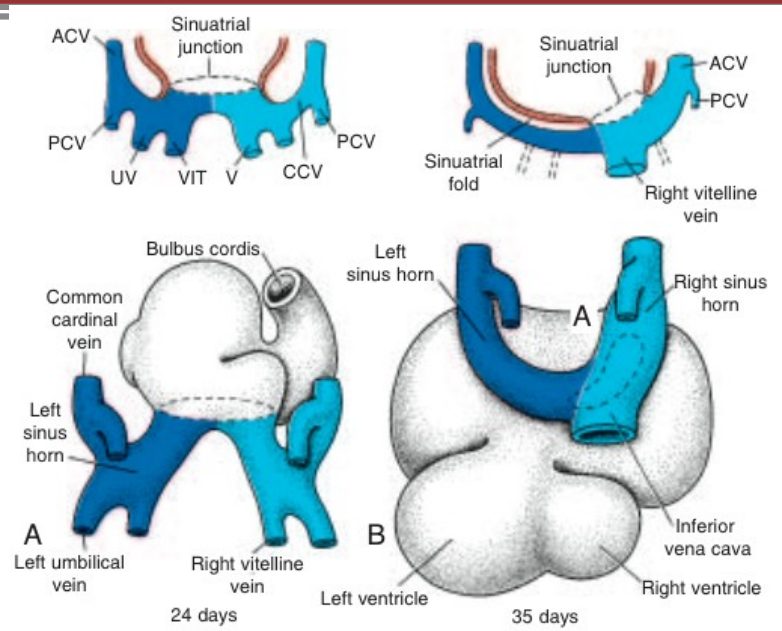
- In middle of **4th week** Sinus venosus receives blood from Rt & Lt sinus horns
- Soon the entrance to atria shifts to right
- Obliteration of
  - Umbilical and left vitelline vein in 5th week and
  - CCV in 10th week

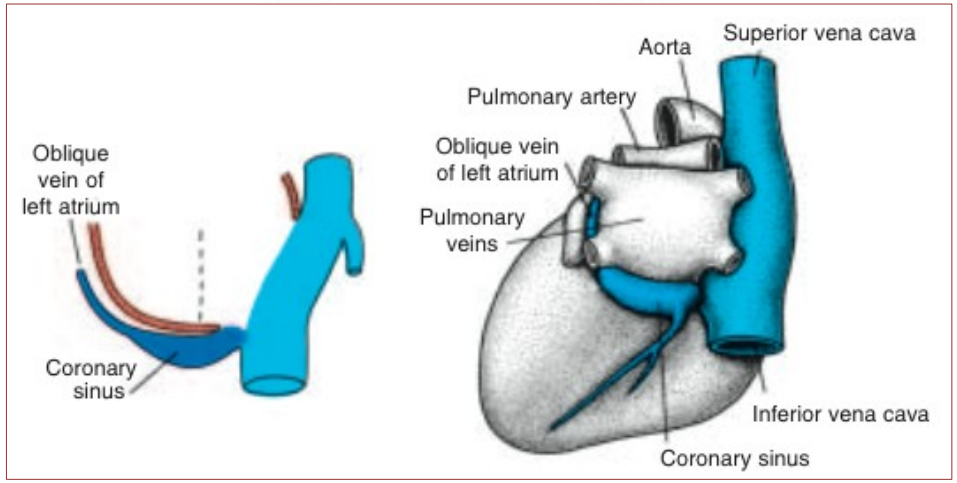


Andersons Paediatric Cardiology, Langman's Medical Embryology 10th Ed

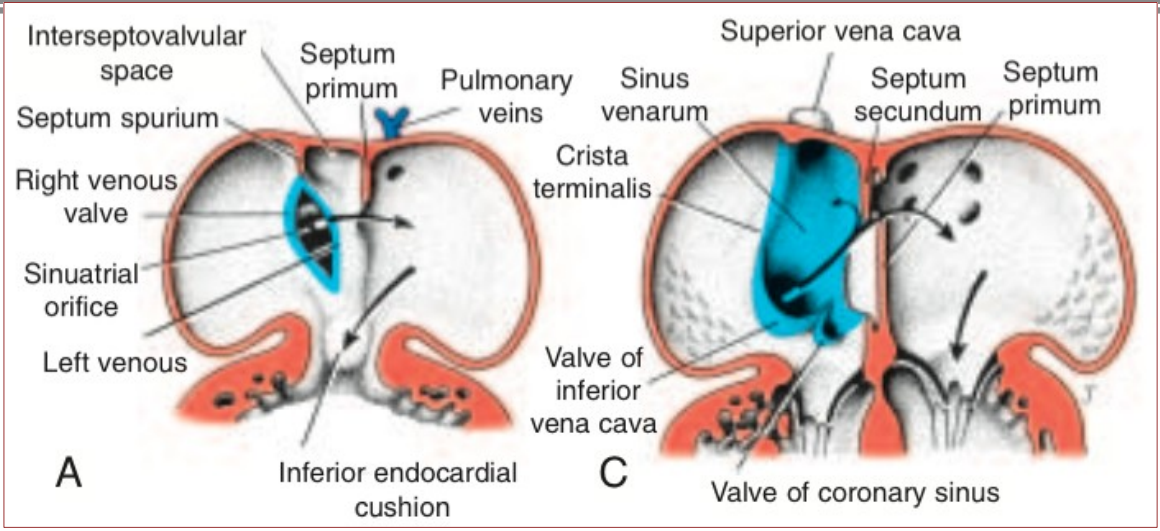
Development of sinus venosus

- Left sinus-
  - oblique vein &
  - coronary sinus





Development of sinus venosus

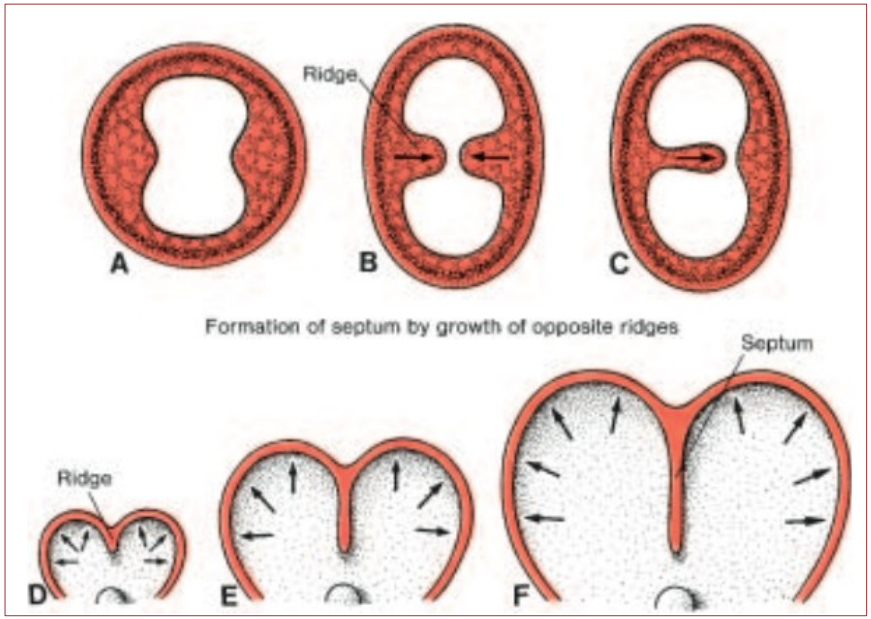


- Sinuatrial orifice is flanked by Rt and Lt venous valves.
- Dorsocranially fused as **Septum spurium.**

- Left venous valve and septum spurium fuse with developing atrial septum.
- Inferior portion of right venous valve: - valve of IVC and valve of coronary sinus
- **Crista terminalis** divides sinus venarum

Formation of cardiac septa

- Major septa formed **27th to 37th day**
  - By growth of single tissue mass- endocardial cushions
  - By expansion of areas on each side of a narrow strip



Andersons Paediatric Cardiology, Langman's Medical Embryology 10th Ed

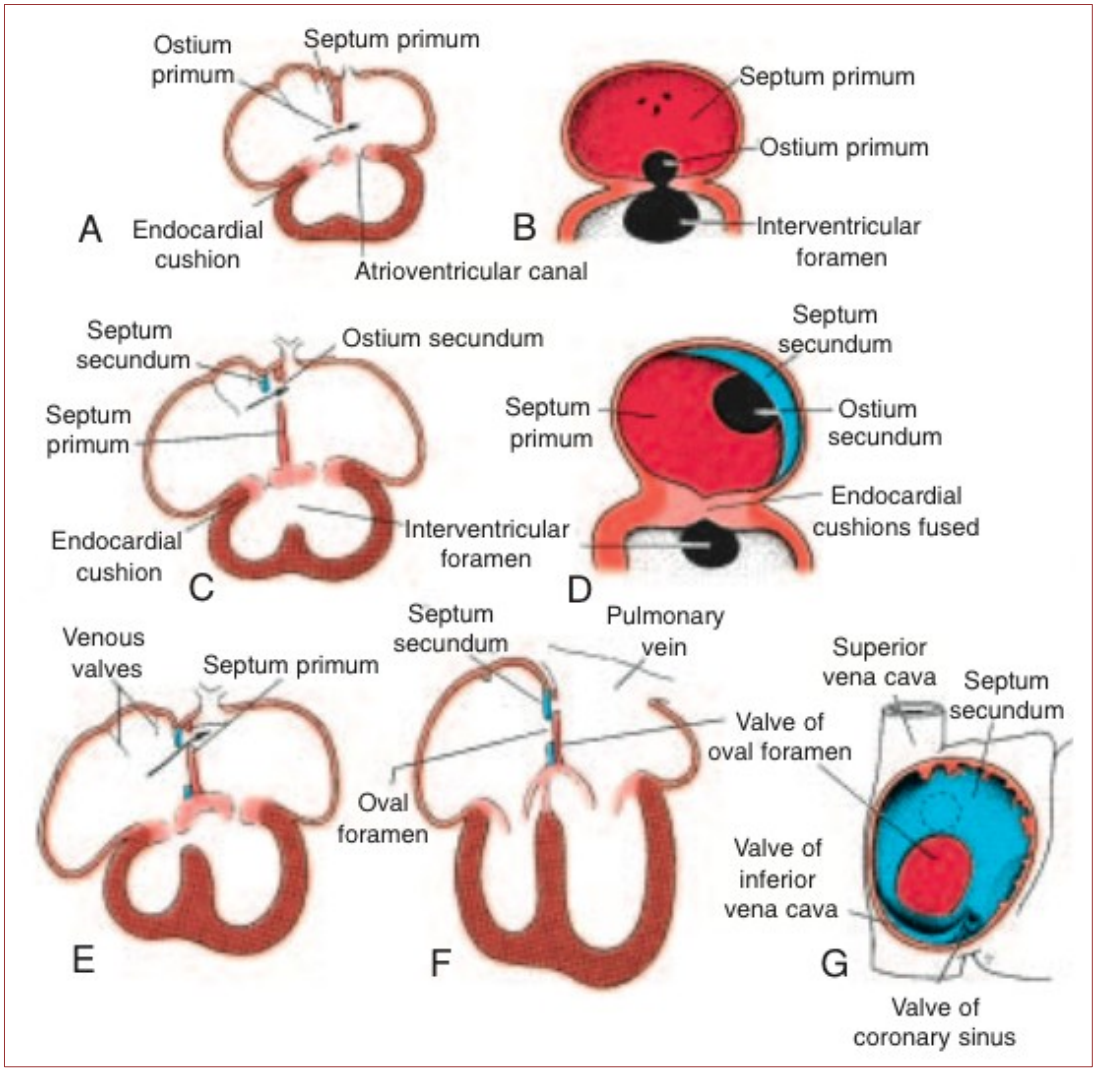
Endocardial cushion defects

- Can lead to ASD, VSD, TGA, TOF
- Cells populating cono-truncal cushions includes neural crest cells
- So often heart and craniofacial defects co-exist

Andersons Paediatric Cardiology,
Langman's Medical Embryology
10th Ed

Atrial septum formation

- At the **end of 4th week**
- Before closure of ostium primum, cell death produces ostium secundum (Morse,1980)
- Opening left by septum secundum is called foramen oval



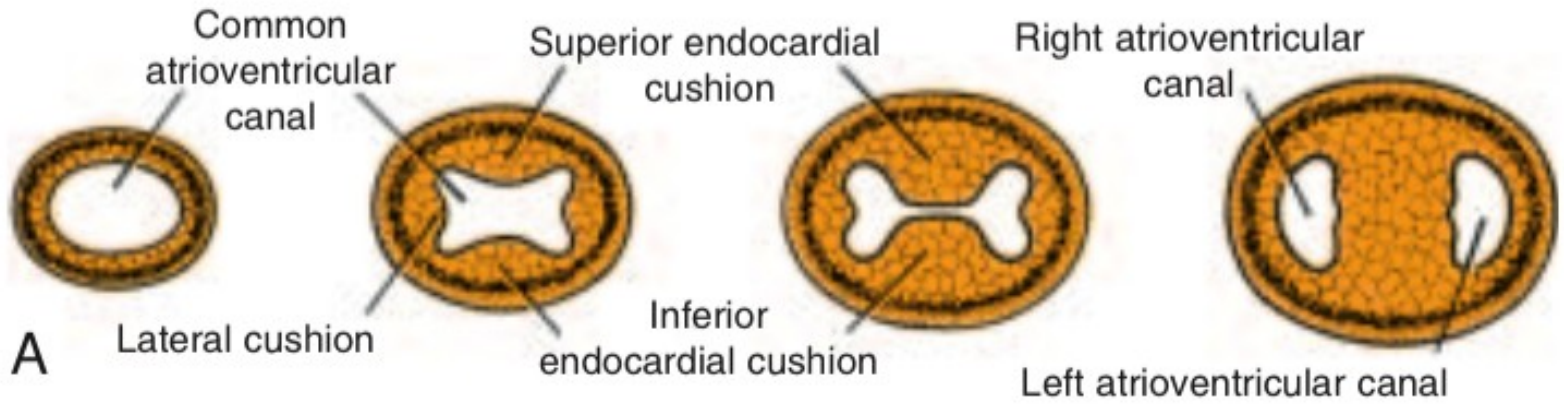
Differentiation of atria

- Pulmonary vein develops as outgrowth from posterior left atrium
- Pulmonary vein and its branches incorporates into left atrium forming large smooth walled part of adult left atrium
- Original embryonic left atrium is little more than the atrial appendage.

- Right horn of CS forms the smooth part of right atrium

Andersons Paediatric Cardiology,
Langman's Medical Embryology
10th Ed

AV canal septal formation.. End of 4th week



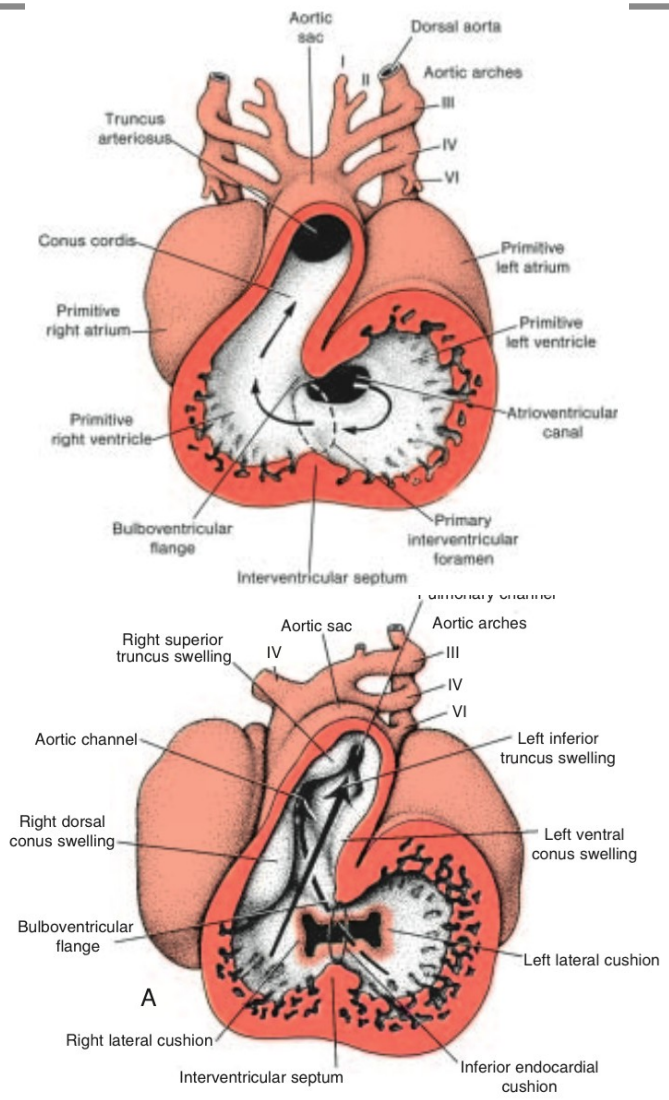
- 2 mesenchymal AV endocardial cushions appear at superior & inferior borders of AV canal
- Later lateral cushions appear

- By the end of 5th week, superior & inferior cushion completely fuse causing complete division of AV canal

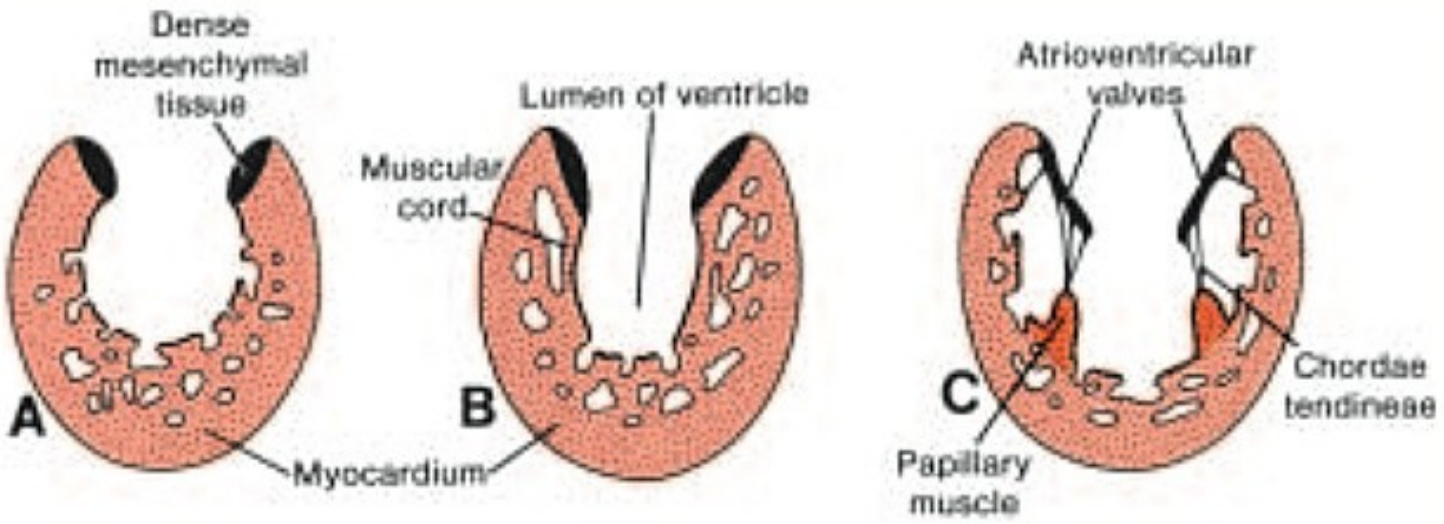
AV canal septal formation

- AV canal separated from bulbous cordis by **bulbo-ventricular flange**
- By end of 5th week, flange is much less prominent and ends midway

- AV canal enlarges
- Blood has access to both ventricles



AV valves



- After fushion of AV cushions- each orifice is covered by local proliferation of mesenchymal tissue.
- Blood flow hollows out and thins out tissue on ventricular surface of these proliferations
- Valves form & remain attached to ventricular wall by muscular cords

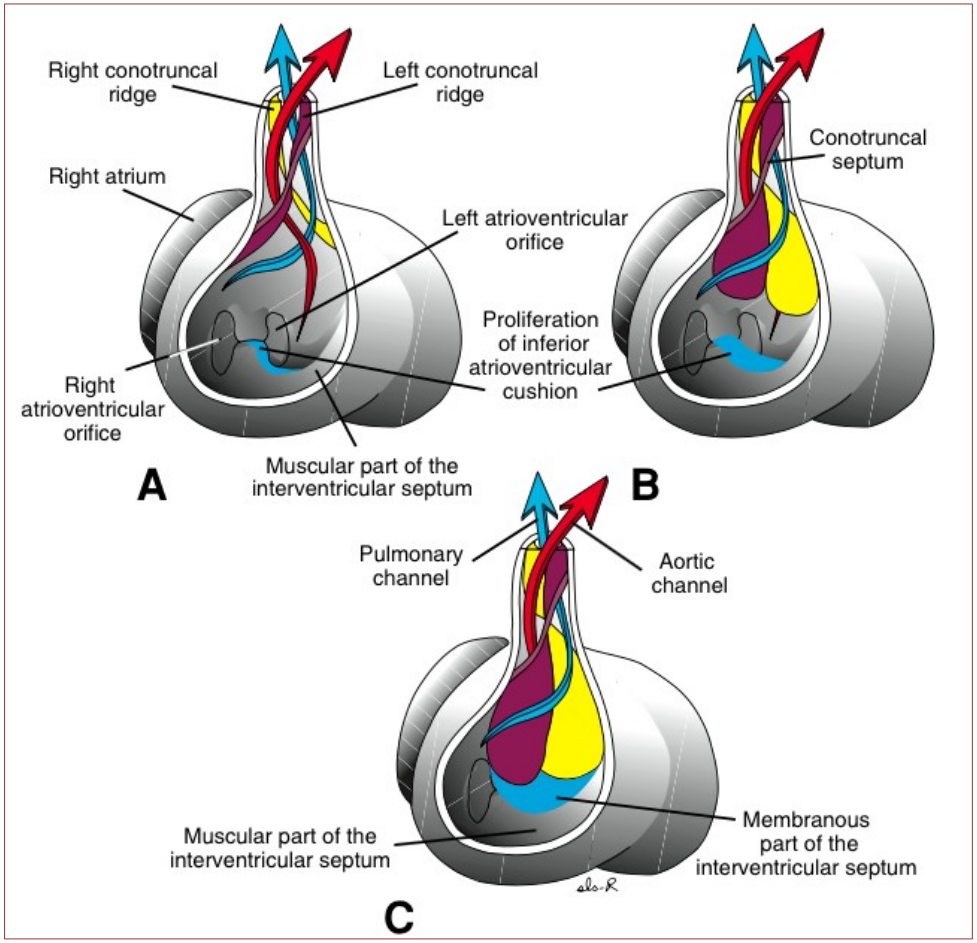
Septum formation in Truncus Arteriosus & Conus Cordis

During the **5th week**

- Pairs of opposing ridges appear in the truncus
- **Right superior** &

Left inferior truncus swelling

- They twist around each other
- After complete fusion



Andersons Paediatric Cardiology, Langman's Medical Embryology 10th Ed

Septum formation in the ventricles

End of 4th week

- Ventricles begin to expand
- Medial walls become opposed and gradually merge forming MUSCULAR septum

- Inter-ventricular foramen - above the muscular portion of the IV septum shrinks on completion of conus septum
- Tissue from inferior endocardial cushion along the top of IV septum- closes the foramen
- Complete closure of this foramen forms the membranous part of IVS

Clinical correlates

VSD:-

(12/10,000)

- Is the most common congenital cardiac malformation
- Involving membranous part (70%) is often associated with conotruncal partitioning anamolies
- Muscular septum VSD in 5 to 20% cases

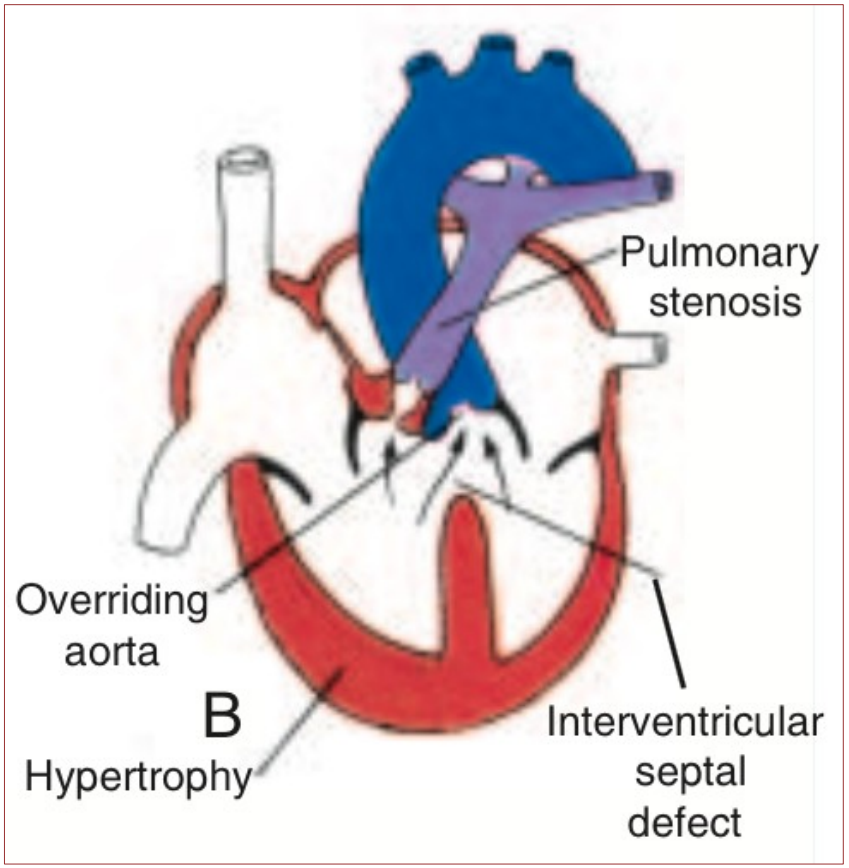
Park's Pediatric Cardiology 6th Ed.

Clinical correlates

TOF:-

(9.6/10,000)

- MC cyanotic CHD
- Most frequently occuring anamoly of conotruncal region
- Results from anterior displacement of conotruncal septum
- 25% have Rt Aortic arch
- 5% have abnormal coronry arteries

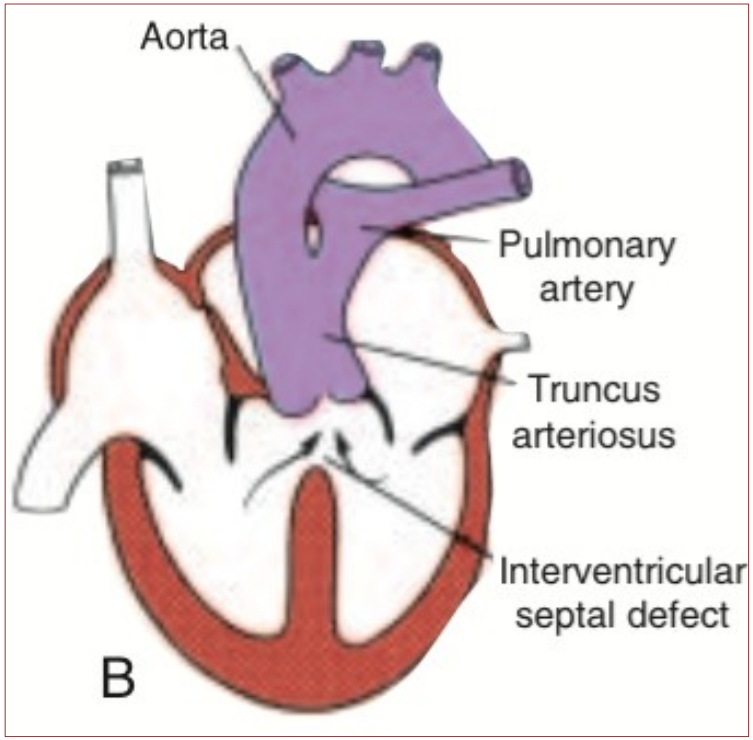


Park's Pediatric Cardiology 6th Ed.

Clinical correlates

Persistant Truncus Arteriosus:- (0.8/10,000)

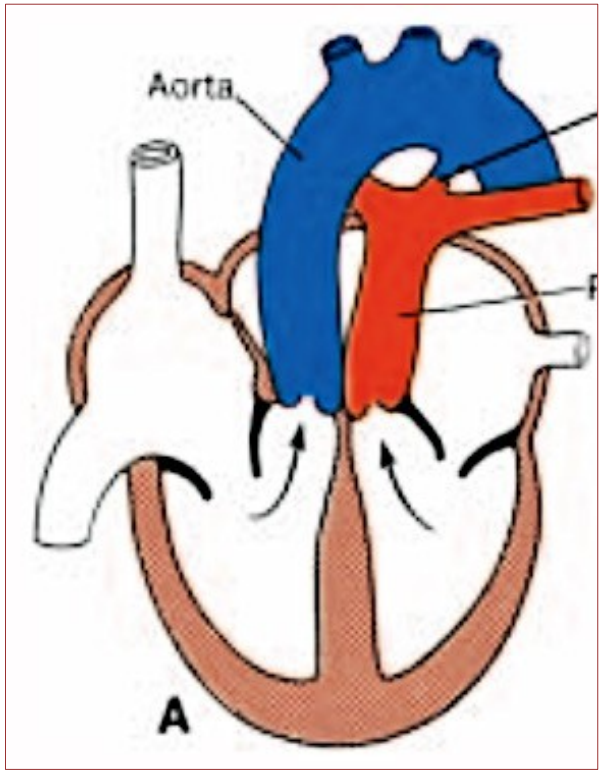
- Conotruncal septum fails to fuse & descend towards ventricles
- Pulmonary artery arises some distance above the origin of undivided truncus
- Ridges also participate in IVS formation
- Always associated with a VSD



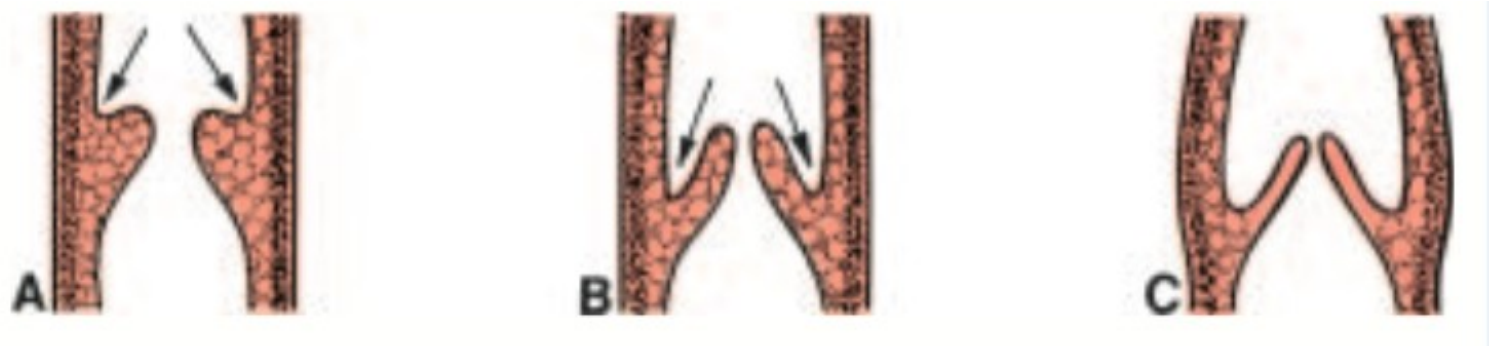
Clinical correlates

Transposition of great vessels (4.8/10,000)

- Conotruncal septum runs straight down instead of spiral
- Usually a/w PDA
- Sometimes a/w Membranous VSD
- Neural crest cells involved
- DiGeorge syndrome:- facial defects, thymic hypoplasia, PTH dysfunction, outflow tract abnormalities- PTA, TOF



Semilunar valves



- Primordia visible as small tubercles on the main truncus swellings
- Gradually the tubercles hollow out at their upper surfaces- forming semilunar valves
- Neural crest cells contribute to formation of these valves

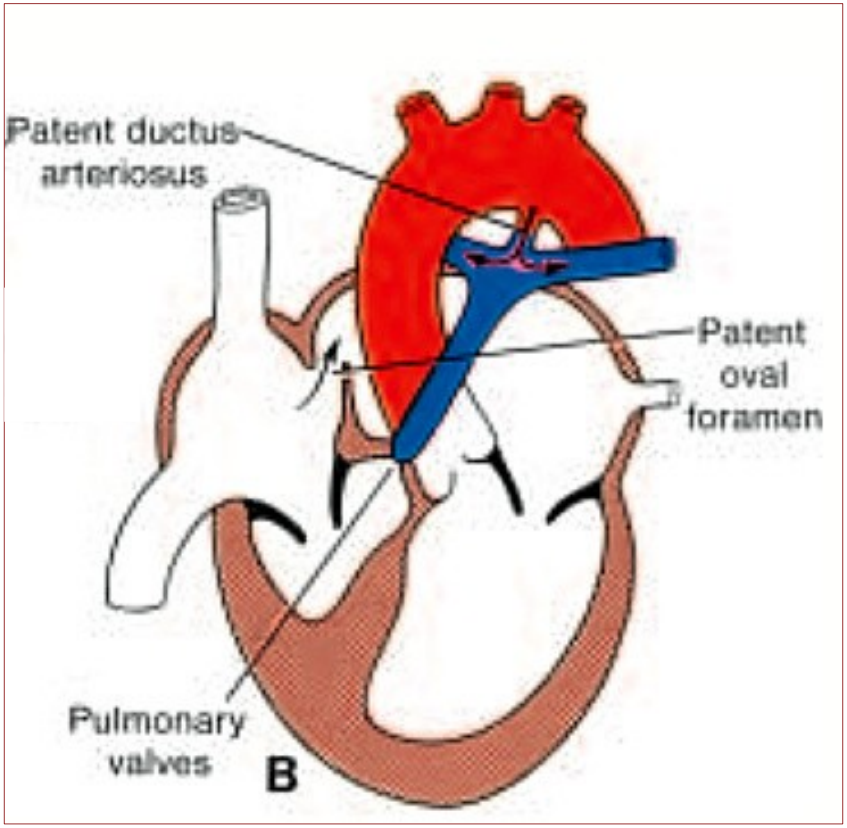
Andersons Paediatric Cardiology, Langman's Medical Embryology 10th Ed

Clinical correlates

Valvular stenosis :-

(3 to 4/ 10,000 births)

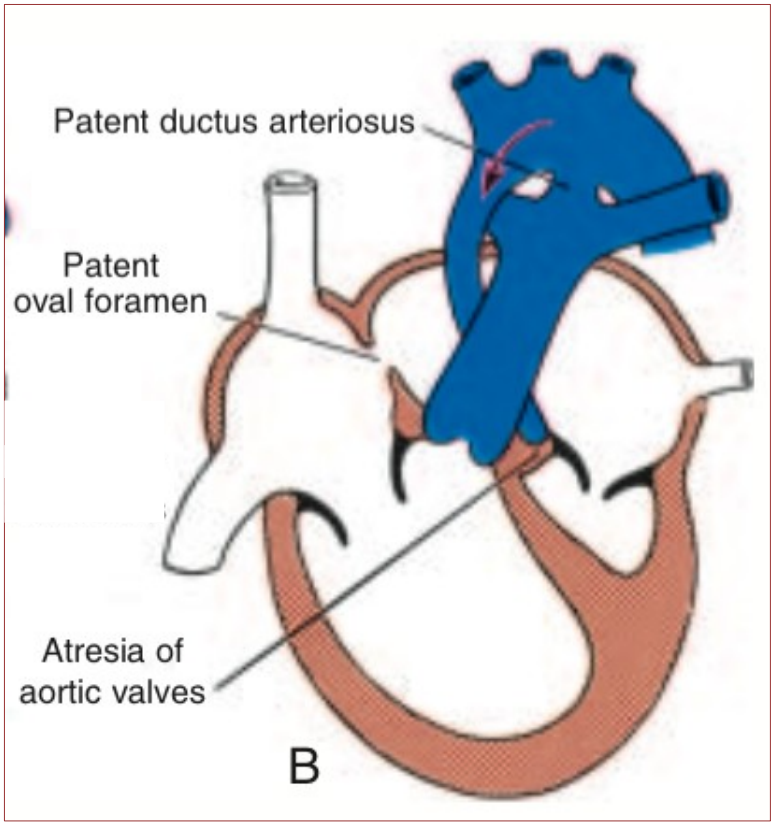
- Valves fused for variable distance
- Trunk of PA is narrow/atretic
- Patent Foramen Ovale- only outlet for blood from right side of heart.
- PDA always patent- only access to



Clinical correlates

Aortic valvular atresia:-

- Fusion of the aortic valves is complete
- Aorta, LV & LA are underdevelopd.
- Usually accompanied with a PDA which delivers blood to the aorta



Formation of conducting system of heart

PACEMAKER:-

1. Pacemaker is initially in caudal part of left cardiac tube
2. Moves to sinus venosus
3. As sinus is incorporated into RA- moves to opening of SVC

Mature conduction system forms from 4 rings of specialized myocardium intervening between primitive segments (Wenink, 1976)

Andersons Paediatric Cardiology,
Langman's Medical Embryology
10th Ed

Formation of conducting system of heart

- The sinuatrial ring-
  - Sinus node
  - Sinuatrial septum- brings the sinuatrial ring to form Av node
  - Internodal pathways (Thorel, James)
- AV ring & primary ring- Entire AV node and bundle (Lamer's et al)

THANK YOU

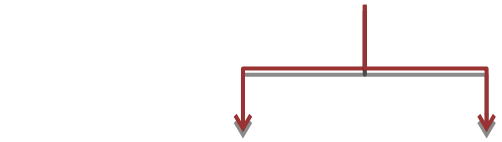
EMBRYOLOGY OF HEART-2

ANOMALIES OF PULMONARY VEINS

Dr. Anurag Bahekar

Headings covered

- Embryology of pulmonary venous system
  - Normal
  - TAPVC
  - PAPVC
  - Cor-triatriatum
- Classification of Pulmonary Venous anomalies
- TAPVC/D
  - Anatomy
  - Physiology



TAPVC without obstruction
- Clinical feature
- Diagnosis

TAPVC with obstruction
- Clinical feature
- Diagnosis

TAPVC/D
- DD
- Treatment
- Prognosis

Embryology

VENOUS SYSTEM OF THE EMBRYO

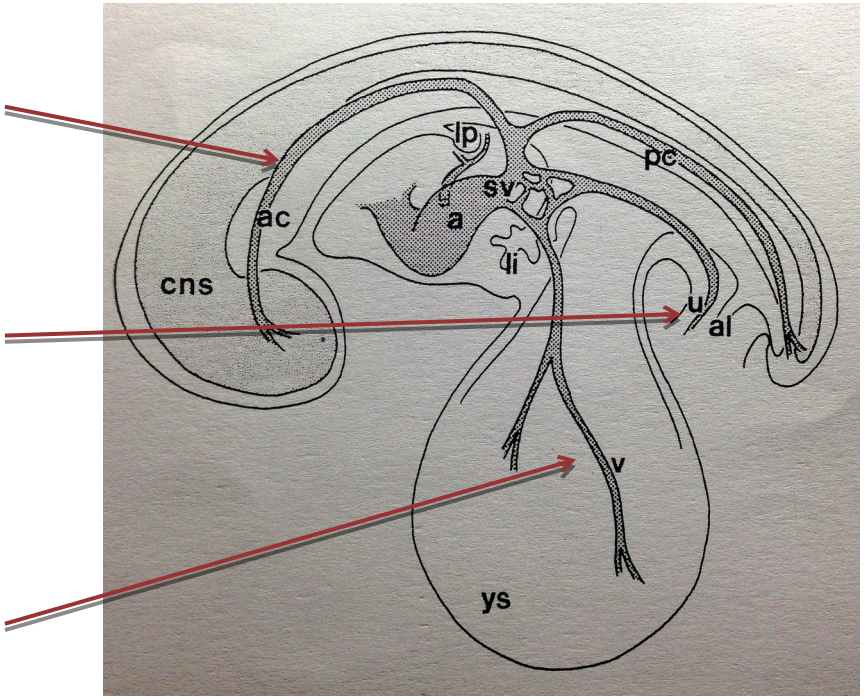
- CARDINAL VEINS-

  These veins drain the body of the embryo proper.

- UMBILICAL VEINS-

  Carrying oxygenated blood from chorionic villi to the embryo.

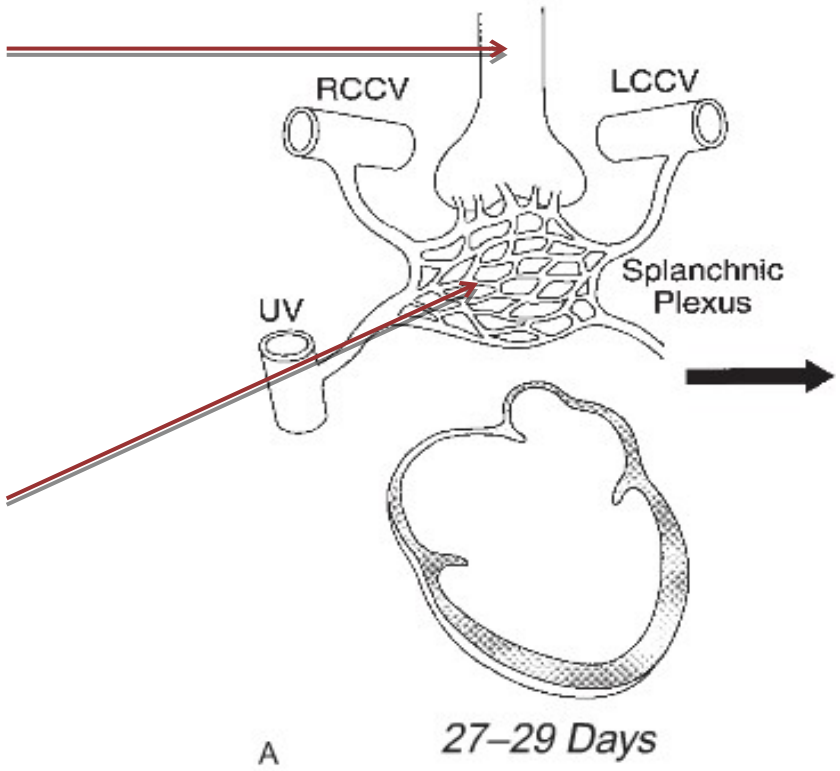
- VITELLINE VEINS (OMPHALOMESENTERIC VEINS)-



Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

Embryology

- In the embryo, the primordia of the lungs, larynx and tracheobronchial tree are derived from a division of the foregut
- Lungs are enmeshed by the vascular plexus of the foregut (splanchnic plexus).
- There are multiple connections with the splanchnic plexus

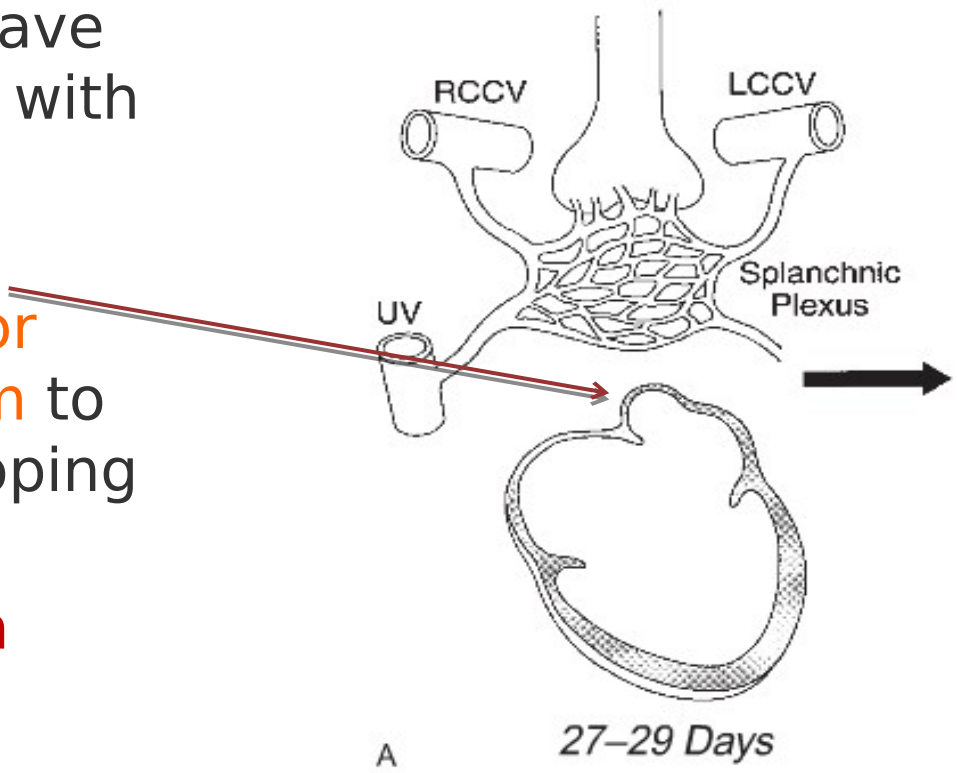


Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

Embryology 27-29 days

Site of development of common pulmonary vein

- At this stage lungs have no direct connection with the heart.
- A small evagination arises in the posterior wall of the left atrium to the left of the developing atrial septum.
- It forms the common pulmonary vein.



Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

Embryology

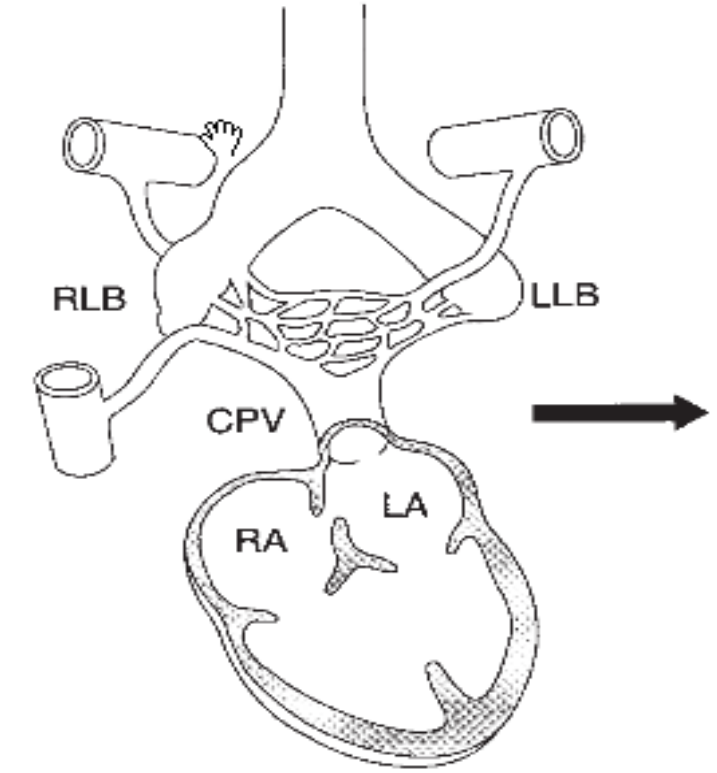
- Incorporation into left atrium of CPV
  (various theories)

1. Evagination in sinuatrial region of heart
2. Starts from confluence of vessels from pulmonary plexus
3. From confluence of capillaries that grow in the mesocardium between lung buds and heart

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

Embryology 32-33 days

- At the end of 1st month
  CPV drains pulmonary plexus into Sinoatrial portion of heart
- Site is cephalad to the junction of Rt & Lt horns of sinus venosus &
- To the left of developing septum primum

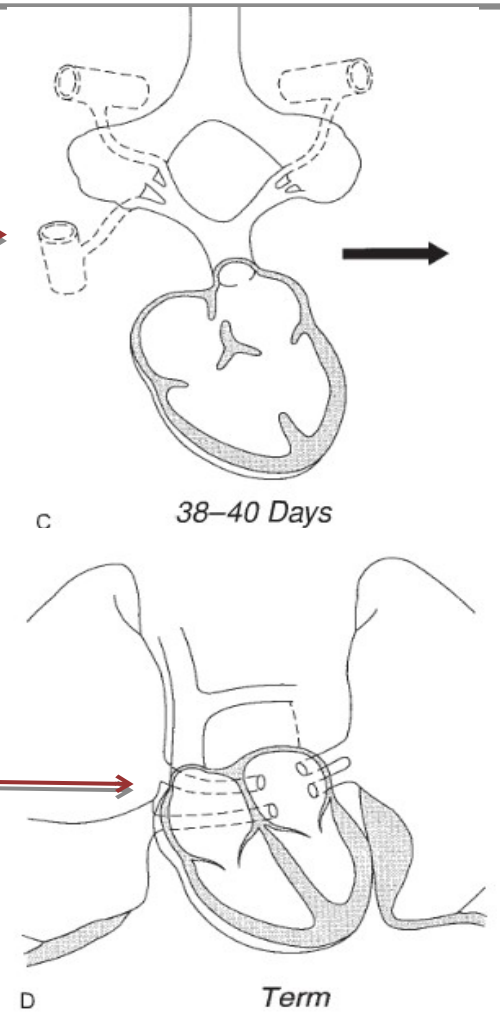


B 32–33 Days

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

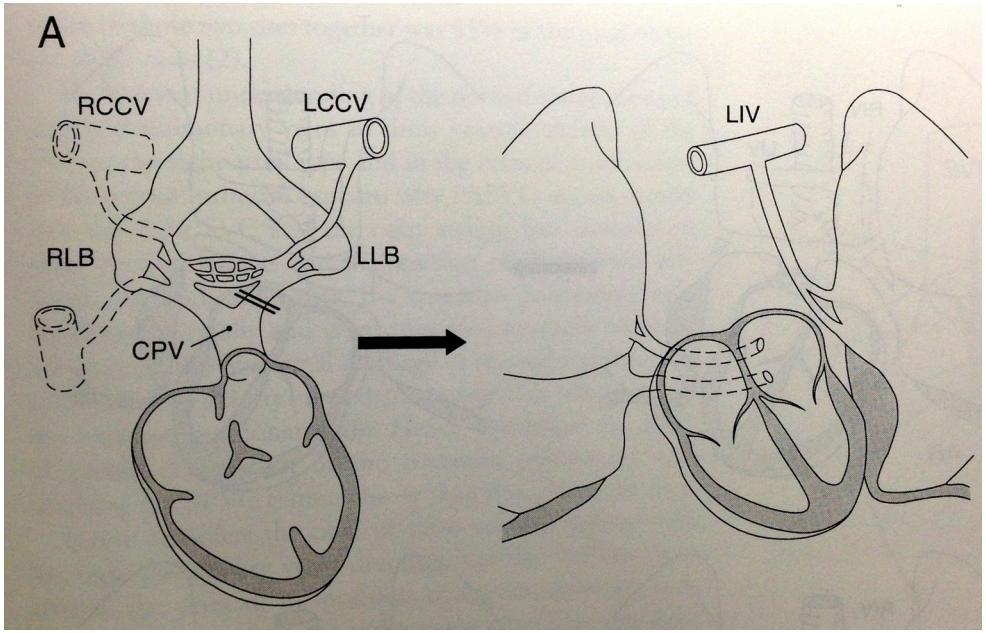
Embryology

- The connection b/w pulmonary venous plexus and splanchnic venous plexus involute
- Common pulmonary vein incorporates into the left atrium
- Individual pulmonary veins connect separately and directly to the left atrium.



Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

Embryology

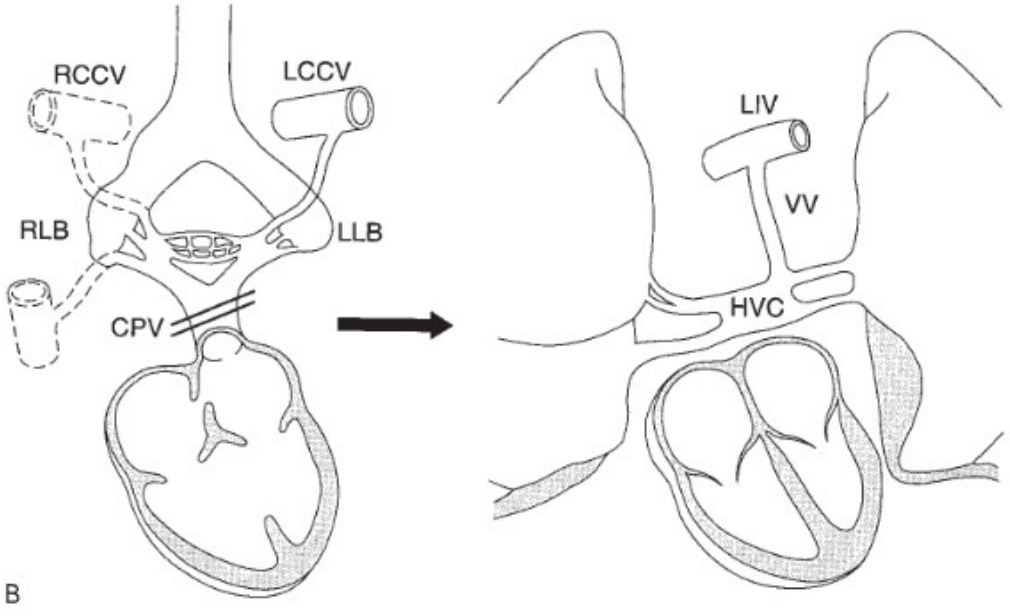


- **PAPVC**

Results due to failure to establish a normal connection between 1/more of pulmonary veins with CPV before the connections with splanchnic venous system have regressed

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

Embryology

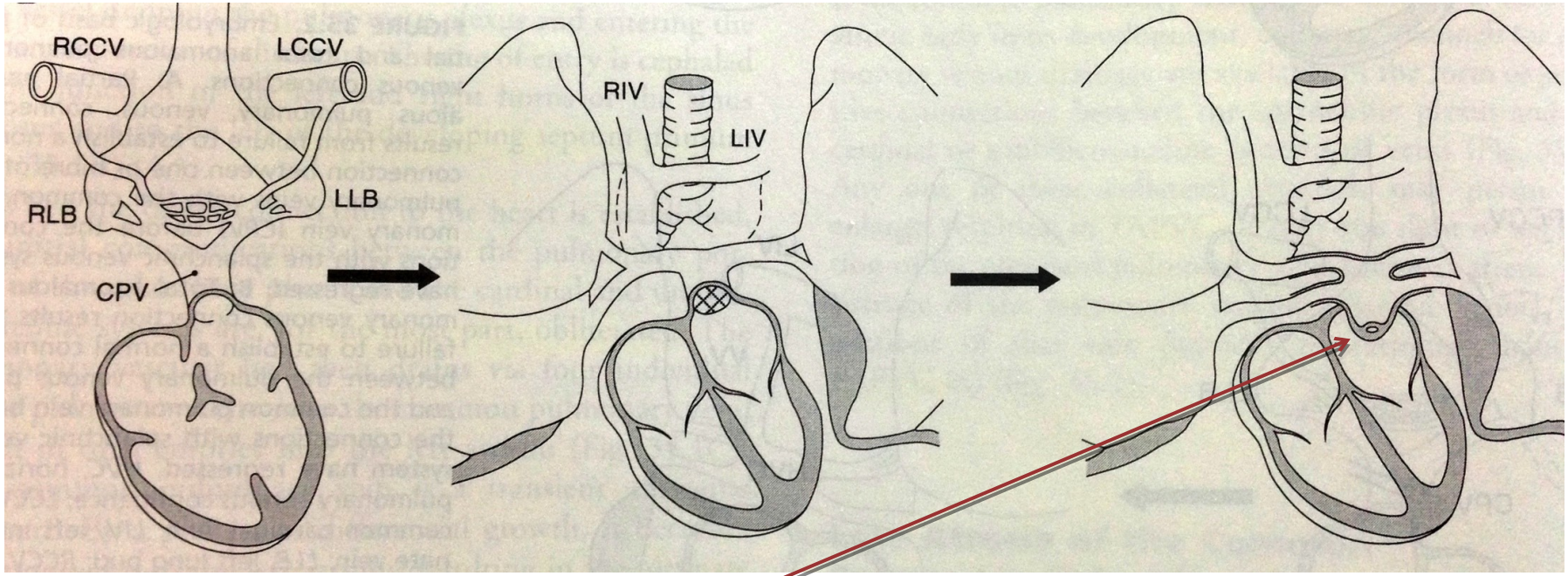


- **TAPVC**

Results due to early atresia of CPV while Pulmonary-systemic connections are still present

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

Embryology



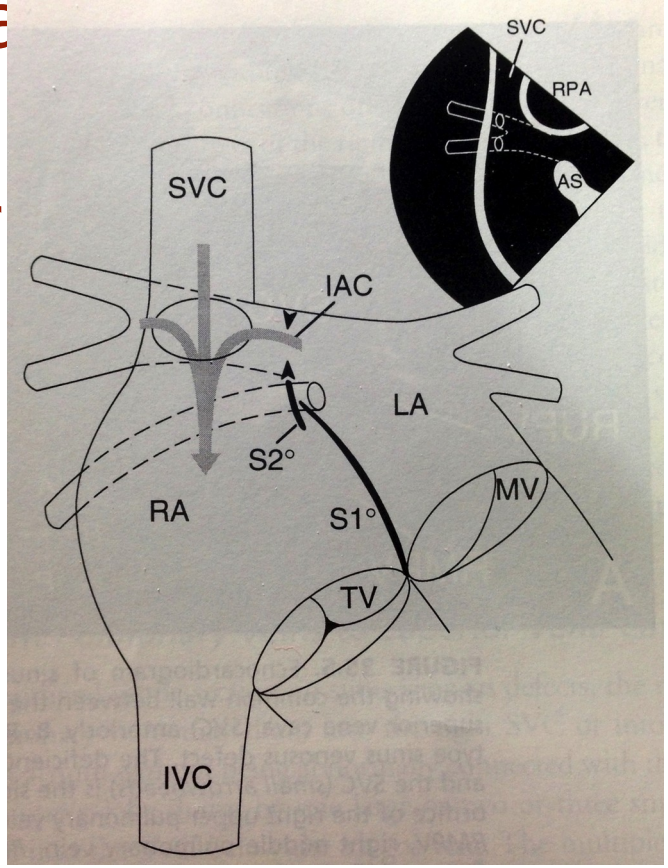
- **Cor-triatriatum**

Stenosis of CPV results in dilatation of the CPV and development of Cor triatriatum

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

Embryology

- Normal absorption of CPV with partially/totally abnorma
  - Sinus Venosus defect
  - Malposition of septum pr



Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

Embryologic Classification Of Pulmonary Venous Anomalies

TABLE 35.1. EMBRYOLOGIC CLASSIFICATION OF PULMONARY VENOUS ANOMALIES

I Normal absorption of the common pulmonary vein associated with defects that result in abnormal pulmonary venous drainage
- A. Sinus venosus defect
- B. Malposition of septum primum

II Atresia of the common pulmonary vein (early) while pulmonary-to-systemic venous connections are still present
- A. Partial anomalous pulmonary venous connection
- B. Total anomalous pulmonary venous connection
  1. Without pulmonary venous obstruction
  2. With pulmonary venous obstruction

III Atresia of the common pulmonary vein (late) after pulmonary-to-systemic venous connections are obliterated
- A. Atresia of the common pulmonary vein

IV Stenosis of the common pulmonary vein
- A. Cor triatriatum

V Abnormal absorption of the common pulmonary vein into the left atrium
- A. Stenosis of the individual pulmonary veins
- B. Abnormal number of pulmonary veins

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC

- Anomaly in which the pulmonary veins have no connection with the left atrium
- Rather, the pulmonary veins connect directly to one of the systemic veins (TAPVC) or
- Drain into the right atrium (TAPVD)

  - WILSON- 1st description in 1798

  "A description of a very unusual formation of the heart",

  Wilson et.al.Philos.Trans.R.Soc.Lon.88;346,1798

  - MULLER- 1st successful open repair in 1951

TAPVC- Genetics and epidemiology

- Mechanism of transmission of TAPVC has not been elucidated
- The Baltimore- Washington Infant study showed a possible association with exposure to lead, paint stripping chemicals and pesticides
- A monogenic pattern of inheritance has been suggested
- Associated with asplenia, polysplenia, cat's eye syndrome

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- Incidence

- In Abbott’s series :- 4/1000 cases
- In another study :- 2% of autopsied cases of CHD

- Marked male preponderance in TAPVC to portal vein.
- M=F in other sites

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- Anatomy

Darling and associates (1957)

1. **Type I-Supracardiac connections :** The common venous confluence joins SVC
2. **Type II-Cardiac TAPVC :** CVC drains into coronary sinus
3. **Type III-Infracardiac TAPVC:** CVC drains into hepatic vein, ductus venosus, portal vein or IVC. It penetrates the diaphragm through the esophageal hiatus.
4. **Type IV-Mixed Type:** at 2/more levels

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- Anatomy

Burroughs and Edwards (with prognostic implications)

- Based on length of anomalous channel
  - Long
  - Intermediate
  - Short

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- Anatomy

Smith et al.

- Supra-diaphragmatic (without pulmonary venous obstruction)
- Infra-diaphragmatic (with pulmonary venous obstruction)

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- Anatomy

- Frequencies of Various types

TABLE 35.2. COMPARISON OF ANATOMIC SITE OF CONNECTION OF TAPVC AND TAPVD IN THREE AUTOPSY SERIES

Site of Connection or Drainage	Burroughs and Edwards (59) (N = 113)	Lucas et al. (60) (N = 71)	Delisle et al. (70) (N = 93)
Left innominate vein	36%	37%	26%
Coronary sinus	16	16	18
Right atrium	15	2	8
Right superior vena cava	11	12	15
Portal system	13	23	24
Multiple sites	7	10	5
Unknown or other	2	0	4

TAPVC, total anomalous pulmonary venous connection; TAPVD, total anomalous pulmonary venous drainage.

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- Anatomy

Connection to RSVC/ RT. Azygous vein

- Pulmonary veins join to form confluence posterior to LA
- Anomalous vessel from Rt side, ascends anterior to Rt lung hilum
- Enters posterior aspect of SVC

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- Anatomy

Connection to lt cardinal system

1. LIV (MC site)

Pulmonary veins form confluence post to LA

↓

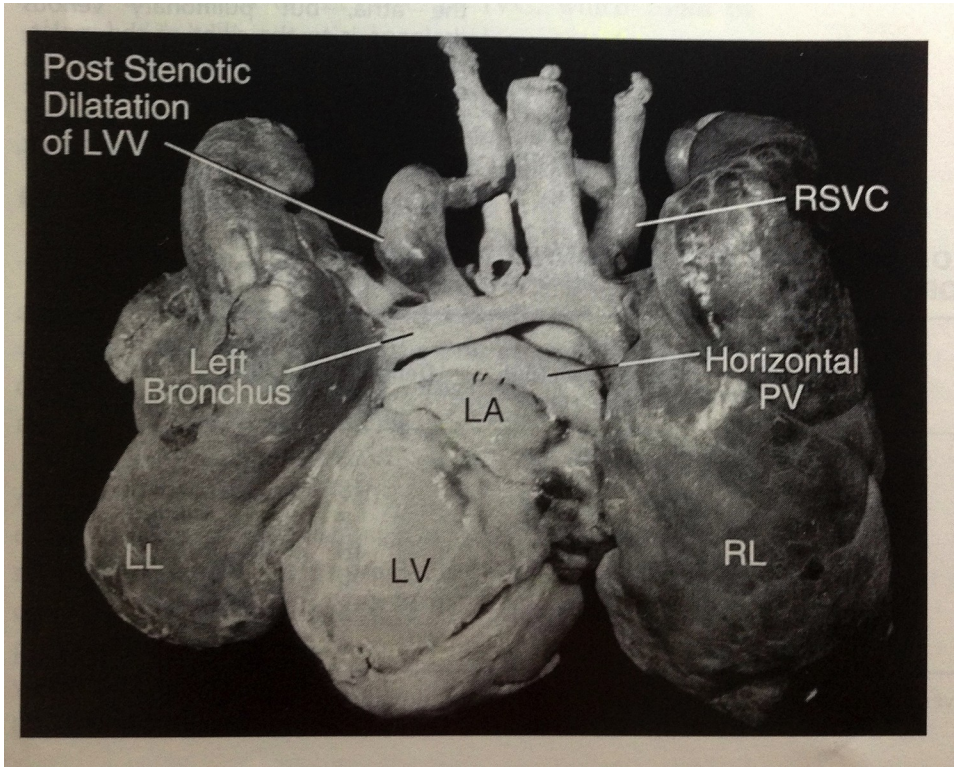
Venous vessel from left side (VV)

↓

Passes ant to Lt pulmonary artery, bronchus, aortic arch

↓

Joins LIV



Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- Anatomy

Connection to lt cardinal system

2. **Coronary sinus**

Pulmonary veins join a common vessel

↓

Connect to coronary sinus

↓

Coronary sinus opens in RA

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- Anatomy

Connection to umbilico-vitelline system

Common confluence immediately behind LA

↓

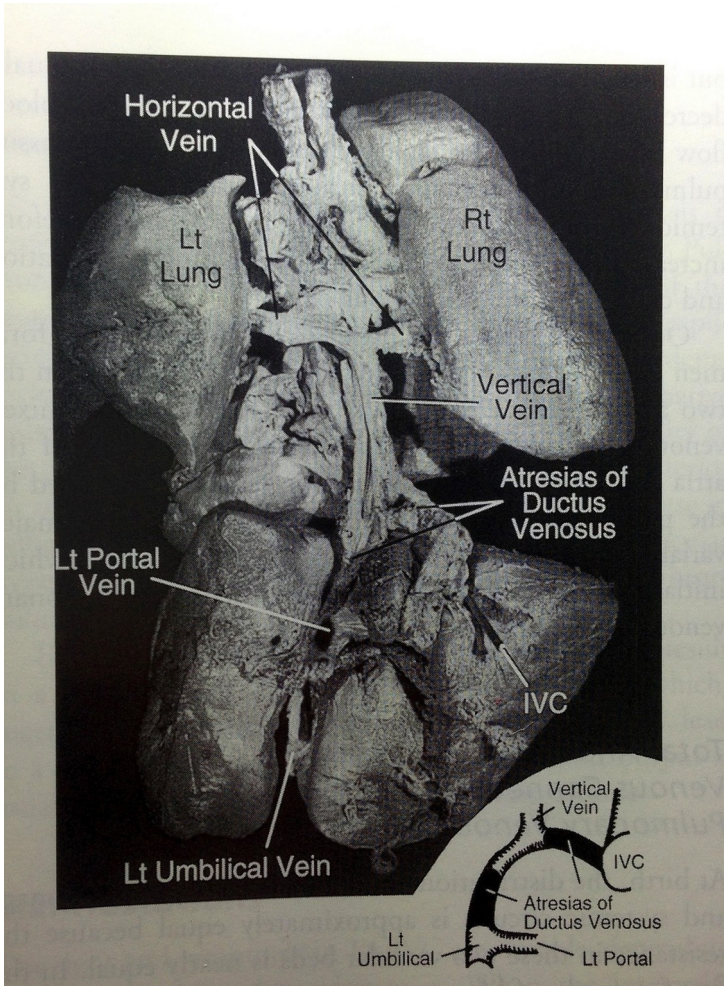
Descends anterior to esophagus

↓

Penetrates diaphragm through the esophageal hiatus

↓

Joins Portal vein at the confluence of splenic & superior mesentric veins



Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- anatomic sites of obstruction

At inter-atrial septum

- Longevity in TAPVC is related to the size of the ASD (Burroughs & Edwards)
- Patients with large defects survived longer

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- anatomic sites of obstruction

In anomalous venous channel

- Intrinsic narrowing is frequent in the walls of anomalous venous channels.
- Extrinsic venous compression may also occur.
- Length of the pathway also offers impedance
- **Infracardiac type is usually obstructive while**
- **Supracardiac and cardiac type are often nonobstructive.**

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- Associated anomalies

- TGA
- TOF
- Single ventricle
- Truncus arteriosus
- Tricuspid atresia
- HLHS
- CoA
- Asplenia or polysplenia

TAPVD to rt atrium – as a rule in patients with visceral heterotaxy and polysplenia

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- Physiology

- All venous blood returns to the RA
- Survival is dependent on presence of a right to left intracardiac shunt (PFO or ASD)
- Physiologic features depend on the distribution of mixed venous blood b/w the pulmonary and systemic circulations

- The major hemodynamics depend on-

1. Size of inter-atrial communication
2. Presence or absence of obstruction to PVR and
3. Relative resistance of systemic and pulmonary vascular bed.

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- Physiology

- A restrictive inter-atrial communication, limits blood to LA and reduces systemic output
- After birth PVR decreases –
  - pulmonary circulation increases.
  - Pulmonary and systemic venous blood- to RA
  - increased RA pressure results in elevated pressure & congestion of both venous circuits.
- In presence of wide communication - distribution of mixed venous blood depends on relative compliance of atria & ventricles and the relative resistance of pulmonary & systemic circulations

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- Physiology

TAPVC without Pulmonary Venous Obstruction

- RA receives the entire VR from systemic & pulmonary circuits.
- After birth PVR decreases and RV compliance increases, so more blood comes to RV.
- Pulmonary blood flow is 3-5 times more than systemic.
- It leads to RV failure in neonates and infants.
- Progressive dilation of RV and PA occurs

- Oxygen saturation is equal in all cardiac chambers (85-90%).
- Later on pulmonary vascular disease develops which leads to severe PHT in 3rd to 4th decades

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- Physiology

TAPVC with Pulmonary Venous Obstruction

- Due to obstruction, pulmonary venous pressure increases
- It leads to increased hydrostatic pressure and pulmonary edema
- Reflex pulmonary arteriolar constriction causes PHTN, Rt ventricular hypertension, hypertrophy and ultimately Rt. Heart failure
- Leftward shift of IVS & decreased inflow from LA, causes low systemic output

TAPVC- without obstruction

Clinical Features

- Usually asymptomatic at birth
- 56% symptomatic in 1st month, rest in 1st year
- 75-85 % die by 1 year of age

- Tachypnea, feeding difficulties - initial symptoms - within first few weeks of life
- Infants have recurrent resp. tract infections and failure to thrive.
- Mild cyanosis is present due to adequate mixing of blood.

- Gradually they develop right heart failure and pulmonary arterial hypertension.

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- without obstruction

Clinical Features

- Mild cyanosis with features of CHF
- Prominent parasternal heave

"Multiple cardiac sounds"

- S1 loud,S2 wide fixed split with loud P2. RVS3 present. S4 in older
- ESM 2/6 at upper sternal border (↑pulmonary flow)
- PSM - TR, MDM - increased flow across TV

- LIV connection- innocent venous hum- at base
  - Not louder in diastole
  - Not altered by change in position

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- with obstruction

Clinical Features

- 72% present in first month of life
- Tachypnea, tachycardia and cyanosis usually after 12 hrs.

(D/D Respiratory distress syndrome-symptoms within 12 hours of life)

- marked pulmonary venous congestion and cyanosis because of reduced pulmonary flow.
- Rapid progression to Dyspnea, feeding difficulty, CRF and death

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- with obstruction

Clinical Features

- When infra-diaphragmatic, venous channel traverses the esophageal hiatus
- Feeding, crying and straining cause additional compression that aggravates the dyspnea and cyanosis.

- Cardiovascular findings may be minimal
- Heart not enlarged
- No significant Rt. ventricular heave
- S2 split, P2 accentuated
- Cardiac murmur often absent
- Hepatomegaly almost always present

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC ECG

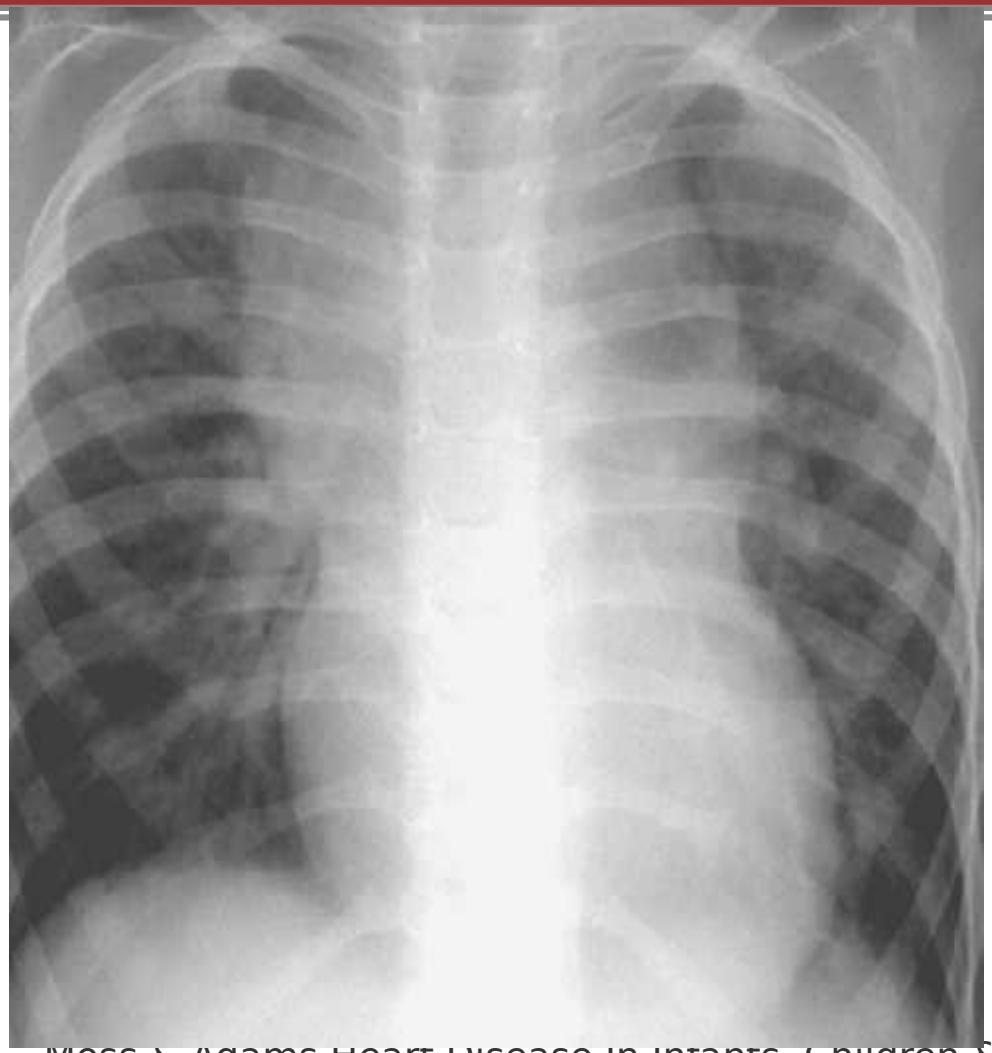
- Tall peaked P wave in lead II characteristic of right atrial enlargement is:-
  - a constant finding of TAPVC without obstruction,
  - NOT in TAPVC with obstruction
- RAD and RVH is usually present (high voltage in right precordial leads).
- Occasionally an incomplete RBBB pattern present.

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- Xray

Figure of 8 or **snow-man** appearance-

- Non-obstructive Supracardiac TAPVC to left innominate vein.
- This diagnostic sign is usually not present in first few months of life.



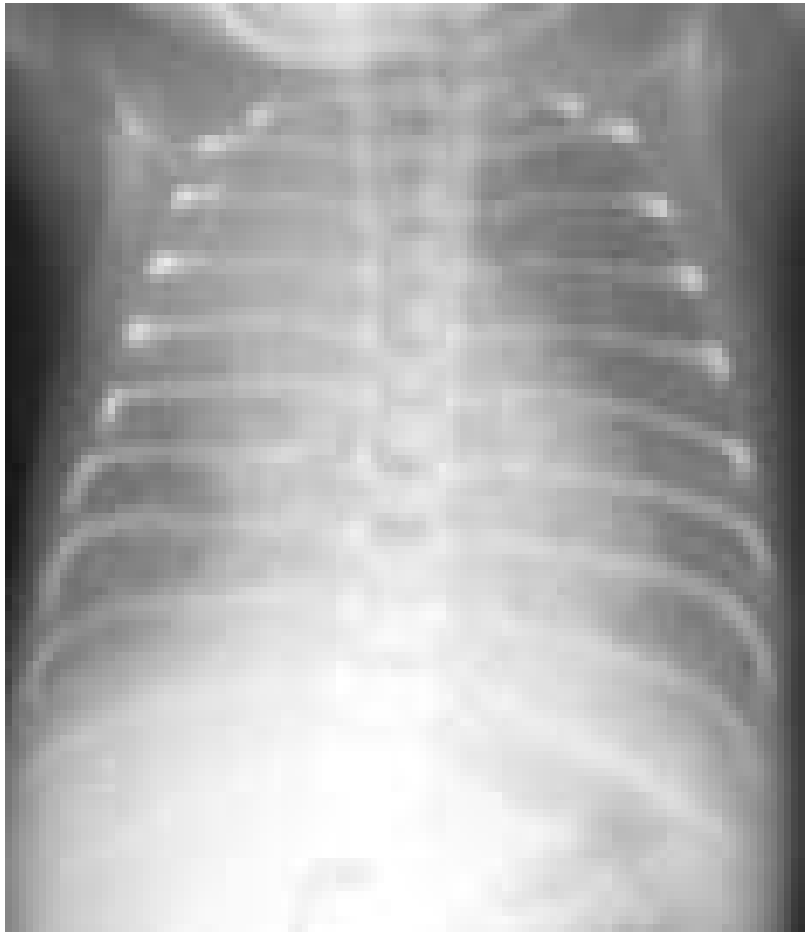
Moss & Adams Heart Disease in Infants, Children & Adolescent 6th Ed.

TAPVC- Xray

Obstructive TAPVC

Ground-glass appearance

- Diffuse reticular pattern
- Cardiac size is normal
- Kerley B lines may be present
- This pattern also seen in other causes of pulmonary venous obstruction.



Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- without obstruction

2D ECHO :- Goals

1. Size of pulmonary veins
2. Connection of all 4 major pulmonary veins to confluence and any additional pulmonary veins.
3. Size of pulmonary venous confluence & its relation with LA.
4. Course of pulmonary venous channel and whether there is obstruction to its flow.
5. To evaluate inter-atrial communication for obstruction.
6. Any additional cardiac anonmaly

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- 2D Echo

- 2D echo with colour Doppler is the definitive non-invasive method for diagnosis of TAPVC.

- The reported sensitivity and specificity of echocardiography was 97% and 99% even before availability of colour doppler.

  Huhta JC et al.Br Heart J 1985;53:525-534

- A sensitivity of 100% and specificity of 85% is claimed for detection of obstruction by 2D echo with colour doppler.

  J Am Coll Cardiol 1991;18:1746-1751

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- 2D Echo

Features common to all forms of TAPVC are-

- Signs of right ventricular volume overload.
- Inability to image the pulmonary veins entering the LA.

- Size of the individual pulmonary vein at the time of diagnosis is a strong, independent predictor of survival.
- Smaller pulmonary veins were associated with poorer prognosis.

Jenkins KJ et al. JACC 1993;22:201-206

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

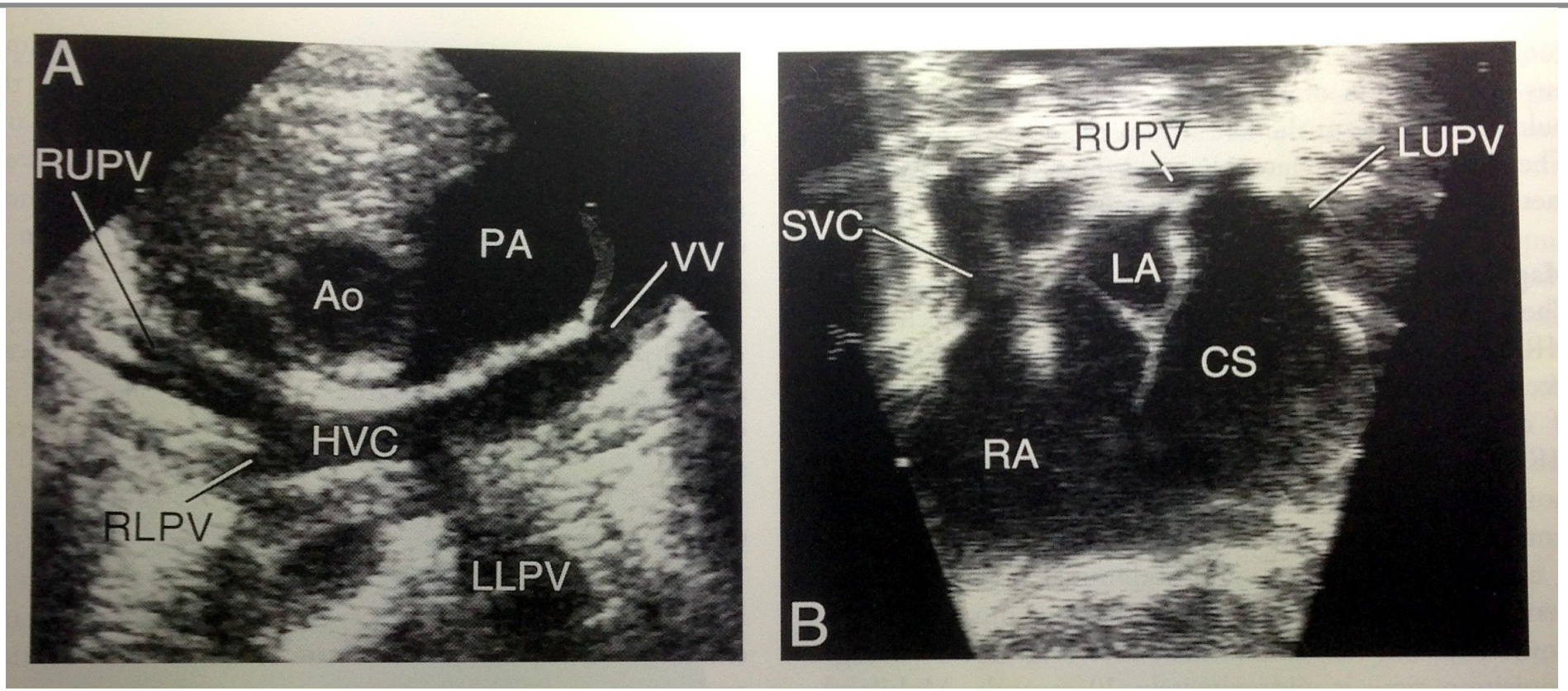
TAPVC- 2DECHO

- Size and orientation (horizontal or vertical) of the pulmonary venous confluence and
- Its relation with the left atrium are important for surgical planning.

- The venous channel in supra-cardiac TAPVC is best imaged from the precordial windows,
- The venous channel in infra-diaphragmatic TAPVC is best evaluated from the subcostal view.

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- 2D ECHO



High lateral Parasternal transverse view TAPVC to LIV

Subcostal oblique view TAPVC to CS

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- 2D ECHO

MC Obstruction Sites

- In supracardiac TAPVC, the site of connection with systemic vein (most frequently the left innominate vein

- In infradiaphragmatic TAPVC, the site of connection with the portal or hepatic vein.

1. Increased flow velocity, (low velocity)
2. Turbulent flow pattern, (phasic laminar pattern)
3. Loss of phasic variations (brief flow reversal during atrial systole) characterize obstructed pulmonary venous flow.

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- 2D ECHO

In TAPVC to coronary sinus,

1. The sinus is dilated,
2. Bulges antero-superiorly into the LA.
3. Imaging of the pulmonary veins draining into the coronary sinus (important because CS may be dilated in other conditions like persistent LSVC to CS)

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- 2D ECHO

Prenatal diagnosis of TAPVC

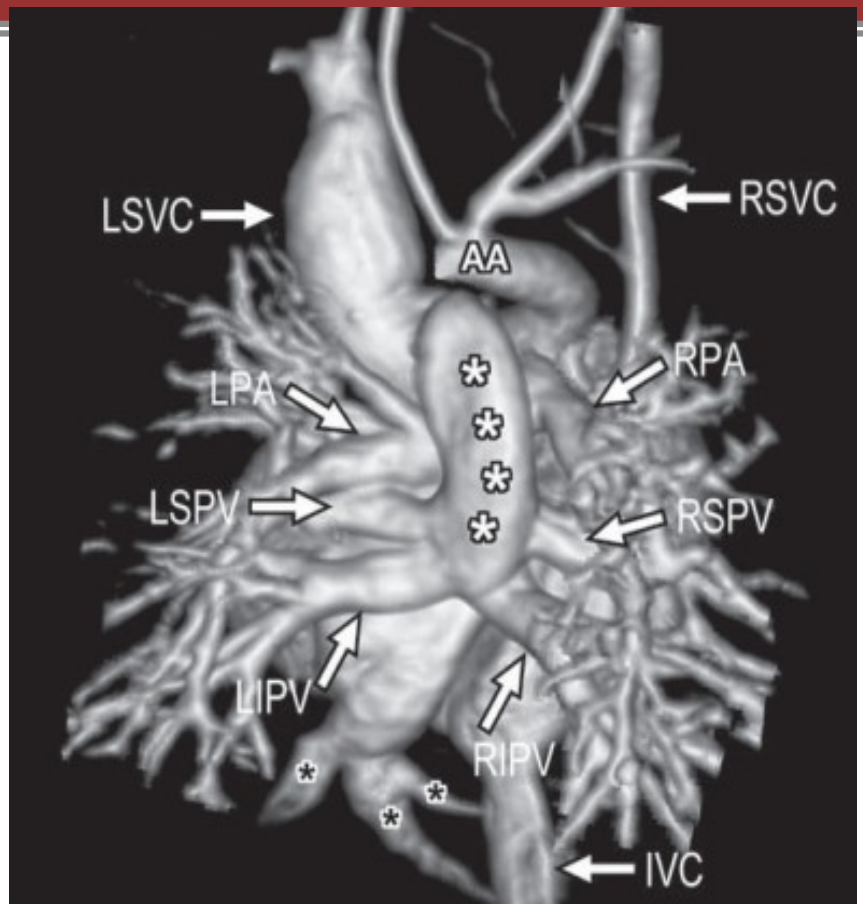
Important for counseling, prognosis and planning for delivery at a center with expertise in pediatric cardiology.

1. As fetal pulmonary blood flow is less in utero, Identification of anomalous pulmonary venous connections is difficult.
2. If RV and PA are enlarged out of proportion to the LV and aorta in utero - then TAPVC is considered
3. (if other causes like CoA, left sided obstructive lesions, AVM are excluded)

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- MRI

- MRI is the preferred imaging technique for evaluation of pulmonary venous structures after echocardiography.
- Lack of ionizing radiation and need for single IV bolus gadolinium contrast are advantages of MRI.
- Long time for acquisition and susceptibility for metal artifact are disadvantages.



Dillman et al.AJR:192,May

TAPVC- Cardiac Catheterization

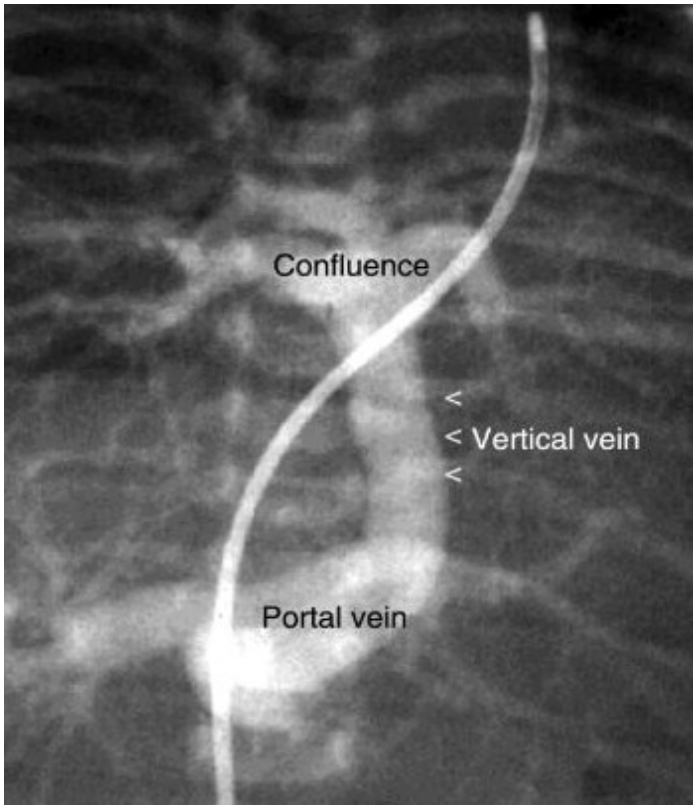
- Rarely performed for TAPVC diagnosis.
- Cardiac cath. is reserved for precise examination of pulmonary veins and their obstruction.
- The pathognomomic finding is oxygen saturation in all chambers and great vessels are nearly identical(80-95%).
- When TAPVC is to left innominate vein or right SVC,
  - SVC blood preferentially flows into tricuspid orifice and
  - IVC blood preferentially shunts into the left atrium,

  resulting in a pulmonary artery O2 saturation that may be higher than that in the systemic artery.

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- Cardiac Catheterisation

- In infracardiac type, anomalous connection of pulmonary veins via descending vertical vein to portal vein is characteristic and it is termed as **TREE IN WINTER**.



Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- Differential Diagnosis

Without obstruction:-

- Large VSD,
- PDA,
- Truncus arteriosus,
- Single ventricle without PS

(Unlike TAPVC)

1. all have xray/ecg- LA &LVH
2. Heart sounds not multiple
3. Harsher murmurs

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- Differential Diagnosis

With obstruction:-

- Hypoplastic left heart
- COA
- Tricuspid atresia
- Pulmonary atresia

(UNLIKE TAPVC)

1. Cardiomegaly
2. Increased arterial vascular markings- TGA, hypoplastic left heart
3. Diminished pulmonary vascularity- Tricuspid atresia, PA

TAPVC- Treatment

Corrective surgery should be performed ASAP

- Mechanical ventilation, inotropic support, diuresis, correction of acidosis & other metabolic problems
- Balloon atrial septostomy/Blade atrial Septostomy:-
  - No longer appropriate
  - Delays definitive procedure
  - Little value if venous channel is obstructed
- Balloon dilatation of obstructed channels- UNSUCCESSFUL (Lock & associates)

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- Treatment

TAPVC to coronary sinus

- First described by Van Praagh et al. in 1972
- Common wall of coronary sinus and left atrium is generously excised through a Right atriotomy approach.
- The enlarged interatrial defect comprising of
  - Coronary sinus orifice
  - Original ASD
  - Intervening excised tissue is closed with a patch

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- Treatment

TAPVC to coronary sinus

- These repairs result in drainage of coronary sinus blood into the left atrium.
- Patients with obstruction in
  - The anomalous pulmonary vein at Vein-Sinus junction or
  - In the coronary sinus have been reportd.
- A wide anastomosis must be made between pulmonary vein and left atrium

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- Treatment

TAPVC to LIV

- Large Side-Side anastomosis between Lt atrium & the CPV (end – side kinks)
- Subsequently the ASD is closed
- Site of anomalous connection is ligated
- Patch is used for ASD closure
- With minor modifications (TAPVC to Rt SVC/ Umbilico-vitelline system)
- Use of stump of amputated LAA- inadequate opening

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- Prognosis

Untreated cases :-

- Factors influencing-
  - Size of ASD
  - Obstructing lesions in anomalous venous pathways
  - State of pulmonary vascular bed

Survey by Keith et.al

- 50% died at 3 mth, 80% at 1 year

In TAPVC with obstruction- oldest survivor 4.5 months

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

TAPVC- Prognosis

Post-operative course

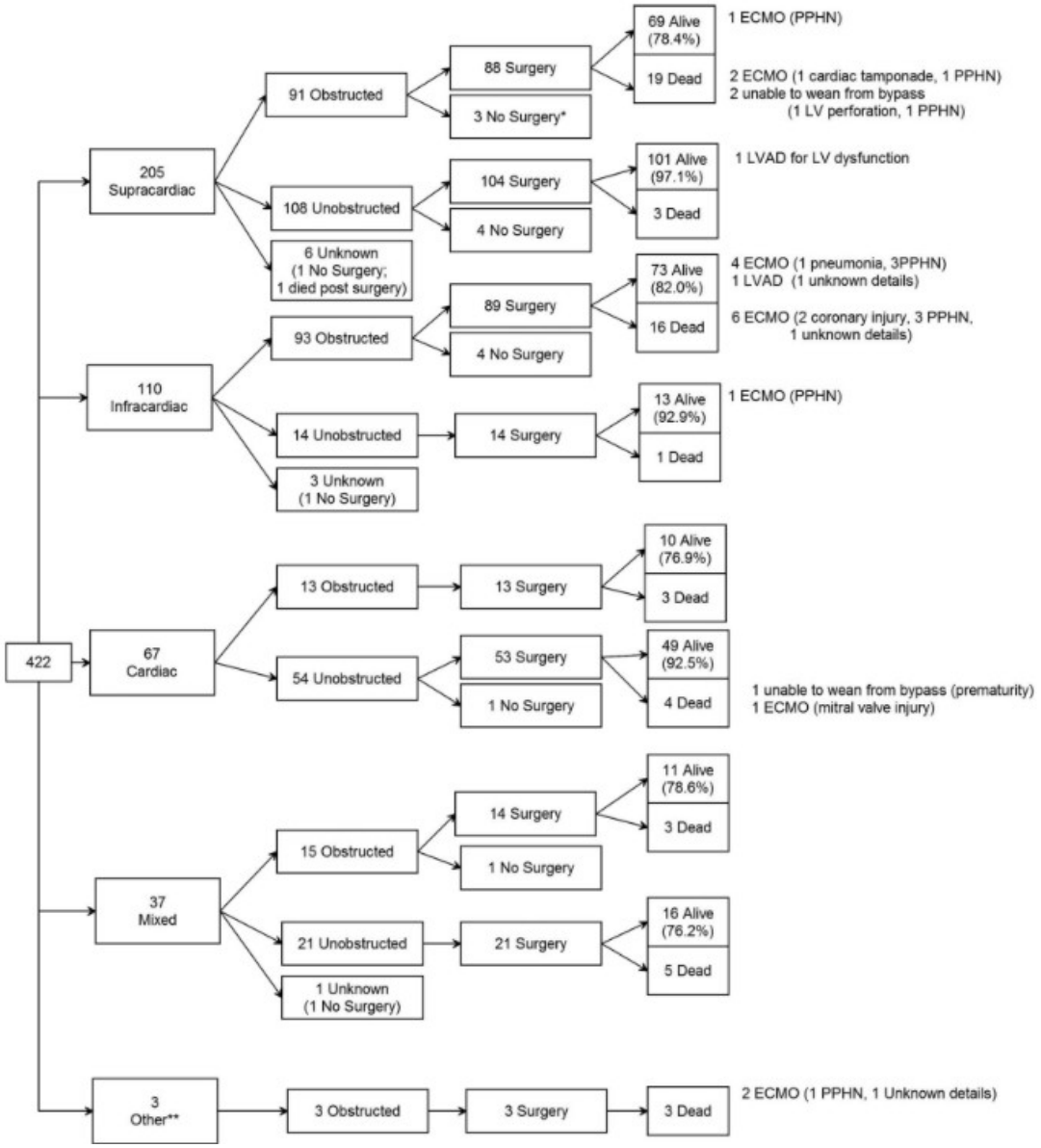
- Long term prognosis appears to depend mainly on
  - State of pulmonary vascular bed at time of operation
  - Adequacy of pulmonary venous- left atrial anastomosis
  - Recurrent pulmonary venous obstruction
- 2D echo with color flow and MRI – useful in evaluating obstruction

- Late arrhythmias-
  - Small no of patients
  - MC- Atrial arrhythmia- sinus bradycardia, Atrial flutter,SVT. VT unusual

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

Review articl

- One of the largest, contemporary, international, population based studies of TAPVC
- Identified risk factors for Postop PVO
  - Mixed cases
  - Hypoplastic pulmonary veins & confluence
- 40% of patients die by 3 yr



Total Anomalous Pulmonary Venous Connection

Morphology and Outcome From an International Population-Based Study

Anna N. Seale, MBBChir, MRCP; Hideki Uemura, MD, MPhil, FRCS;

(Circulation, 2010;122)

Conclusions

- TAPVC is a rare congenital heart anomaly that presents as a surgical emergency in neonatal periods.
- Echocardiography is the diagnostic modality of choice.
- Cardiac catheterization is rarely needed for diagnosis.
- Surgical correction is the definitive treatment.
- Improved surgical techniques and hospital care have led to significantly better outcomes of TAPVC surgery.

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

THANK YOU

EMBRYOLOGY OF HEART-3

ANOMALIES OF PULMONARY VEINS

Dr. Anurag Bahekar

PAPVC- Anatomy

- RPV to SVC
- RPV to IVC
- LPV to IVC
- LPV to LIV
- Other sites

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

PAPVC- Anatomy

- RPV to SVC

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

PAPVC- Anatomy

- RPV to IVC

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

PAPVC- Anatomy

- LPV to IVC

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

PAPVC- Anatomy

- LPV to LIV

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

PAPVC- Anatomy

- Other sites

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

PAPVC- Physiology

- PAPVC with Intact Atrial Septum

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

PAPVC- Physiology

- PAPVC with Atrial Septum Defect

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

PAPVC- Clinical Features

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

PAPVC- Associated defects

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

PAPVC- Diagnostic features

- ECG

PAPVC- Diagnostic features

- Xray

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

PAPVC- Diagnostic features

- Echo

PAPVC- Diagnostic features

- MRI

PAPVC- Diagnostic features

- Cath/ Angiography

PAPVC- Differential diagnosis

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

PAPVC- Treatment

- Medical Management

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

PAPVC- Treatment

Surgical

- PAPVC to RA due to malposition of Septum Primum
- PAPVC to RA due to Sinus Venosus defect
- PAPVC to IVC
- PAPVC to Coronary Sinus
- PAPVC of LPA to IV

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

PAPVC- Surgical Treatment

- PAPVC to RA due to malposition of Septum Primum

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

PAPVC- Surgical Treatment

Surgical

- PAPVC to RA due to Sinus Venosus defect

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

PAPVC- Surgical Treatment

Surgical

- PAPVC to IVC

PAPVC- Surgical Treatment

Surgical

- PAPVC to Coronary Sinus

PAPVC- Surgical Treatment

Surgical

- PAPVC of LPV to IV

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.

PAPVC- Prognosis

Moss & Adams Heart Disease In Infants, Children & Adolescent 6th Ed.